The role of overnight bladder drainage in the children with fulgurated posterior urethral valves with deteriorating renal function and increasing bladder dysfunction.

CERTIFICATE

This is to certify that the dissertation entitled **"The role of overnight bladder drainage in the children with fulgurated posterior urethral valves with deteriorating renal function and increasing bladder dysfunction"** is the bonafide original work of **Dr. Jyotish Kumar** submitted in partial fulfillment of the rules and regulations for the **M. Ch. Branch-V, (Paediatric Surgery), examination of the Tamilnadu Dr. M.G.R. Medical University, Chennai** to be held in **February 2007.**

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INTRODUCTION

Posterior urethral valves are the most common cause of obstructive uropathy involving the lower urinary tract in childhood. Such patients may present from early neonatal period to late childhood with a broad spectrum of clinical severity ranging from poor urinary stream to renal failure. The increased use of antenatal ultrasound has resulted in more patients presenting in the neonatal period. The many options available in the management of the newborn with posterior urethral valves include immediate upper tract diversion, lower tract diversion, valve ablation alone, and valve ablation with immediate upper tract reconstruction. The appropriate procedure must be tailored to the status of the upper tracts.

In many patients, the impairment of renal and bladder function exists despite successful early valve ablation. Increased voiding frequency, small voiding volumes, urinary incontinence and large post-void residuals are frequently observed in these children. In time, myogenic failure may occur with large bladder volumes without sense of bladder fullness and weak detrusor activity leading to valve bladder syndrome. Thus, treatment of these children with poorly compliant bladders usually involves clean intermittent catheterization (CIC) to facilitate bladder emptying and anticholinergic drugs to improve poor bladder compliance. However, in some polyuric cases even catheterization as frequently as every 2 hours is not effective to maintain “safe” bladder

volumes. Furthermore, sustained bladder over distension persists during sleep, and leaves the bladder and upper tracts at risk despite frequent intermittent daytime drainage.

For these cases, one suggested treatment has been the addition of overnight bladder drainage to the prescribed daytime intermittent drainage.

The aim of this study is to assess the role of overnight bladder drainage (OBD) in children with fulgurated posterior urethral valves with deteriorating renal function and increasing bladder dysfunction.

REVIEW OF LITERATURE

Posterior urethral valves are the most common cause of congenital obstructive uropathy in male children, and have variable immediate and long-term effects on the lower and upper urinary tract. The accurate incidence of posterior urethral valves is difficult to ascertain and is approximately 1 in 5000 to 8000 infant males.[1] The urethral obstruction causes damage to the renal parenchyma and damage to the smooth muscle of ureter and bladder. These changes begin prenatal and persist even after successful valve ablation. The spectrum of severity for this disorder ranges from newborns whose conditions are incompatible with life, to patients presenting in late childhood with minimal voiding disorders and normal renal function.[2]

Historical Perspectives and Changing Concepts:

Congenital valvular obstruction of the posterior urethra was first recognized by Young, Frontz, and Baldwin in 1919 and they classified it into three distinct types as follows: a Type I valve is the bicuspid type that usually is seen originating on the floor of the urethra arising from the distal lateral aspect of the verumontanum extending distally and anteriorly to fuse in the midline, Type II valve runs between the verumontanum and the bladder neck, while type III valve appears distal to the verumontanum as a circular, non-oblique diaphragm usually just distal to the verumontanum**.**[3]

But the first generalized recognition of posterior urethral valves became possible only during the era of 1950s and 1960s with the invention of more sophisticated radiological and endoscopic equipments.[4] The mortality rates remained relatively high during this

interval, average exceeding 20% with more than 45% mortality in infants less than 1 month of age.[5]

Awareness of posterior urethral valves became much more widespread among referring physicians during the 1970s, and patients tend to be recognized and referred earlier.[5] The mortality rates improved dramatically during this era. Several authors have repeatedly stressed that in the milder form, posterior urethral valve have a good prognosis[6] but in its most severe form, posterior urethral valve are very serious.[5,7,8,9] It was also noted during this era that the earlier in life renal insufficiency occurs, the more severe the growth impairment is.[10]

During this time, objective criteria evolved for the determination of severity and prognosis of a particular patient with posterior urethral valves: 1) age of presentation.[5] 2) type of presenting problem,[5,8] 3) presenting creatinine;[5,8,10] 4) nadir creatinine;[11]5) presence of unilateral reflux;[12] 6) presence of bilateral reflux;[5] 7) 35 cm of water bladder pressure below bladder capacity;[7,8] 8) presence of bladder diverticulum;[12] 9) severe ureterectasis;[7,8] or 10) ultrasonic demonstration of renal dysplasia.[13]

In the late 1980s and 1990s, just when objective criteria for staging the severity of posterior urethral valves were evolving and more formal and perhaps prospective studies were seriously under consideration, the picture changed completely with the widespread introduction of antenatal diagnosis of urethral valves by ultrasonography.[14]

These developments pointed out the obvious requirement for intrauterine physiologic and pathologic studies. The age of presentation and presenting problems were no longer valid, because the patient was identified in utero by ultrasonographic screening. Previously important criteria such as presenting and nadir creatinine concentrations were

significantly changed. Obviously, in the new era, the presenting creatinine most frequently reflected the mother's creatinine. In addition, a new kind of clinical problem, the potentially lethal case in utero, was identified.[14,15]

Treatment Options: Historical prospectives

Initial attempts at managing urethral valves were described by Young and Frantz in 1919. They used an open suprapubic approach with direct visual destruction of the valves.[3]

During the 1960s, various techniques of high ureteral diversion without tubes were introduced. Johnston, in 1963 introduced temporary cutaneous ureterostomy in the management of advanced congenital urinary obstruction.[16]

Johnston, in 1966 described the first transurethral approach for posterior urethral valve management using an otoscope to locate the valves through a perineal urethrostomy and then hook and destroy them.[17]

Mechanical disruption of urethral valves by retrograde withdrawal of a Foley or Fogarty catheter with the balloon inflated has also proved effective in destroying the obstruction but had a high incidence of extravasations at the site of valve disruption, thus, never been well accepted for fear of creating a urethral stricture.[18,19]

During the 1970s, the introduction of fibreoptic lighting and the Hopkins rod lens system in paediatric endoscopes completely changed the whole course of events making primary ablation of urethral valves possible in almost any infant.

Five schools of operative management of posterior urethral valves evolved:

1. Valve ablation plus serial observation[4]
2. Valve ablation plus immediate reconstruction[20]
3. Valve ablation plus delayed reconstruction[21]

4. Vesicostomy followed by valve ablation and reconstruction[22]
5. High urinary diversion (pyelostomy) followed by valve ablation and reconstruction[10]

Controversy arose and continues as to the optimal therapy.[6,7]

Ehrlich and Shanberg, in 1988 described the use of Nd-YAG laser endoscopically for the management of posterior urethral valves.[23]

Antenatal Diagnosis:

About 10 % of antenatal diagnosed obstructive uropathy is due to posterior urethral valves of which approximately two-thirds are diagnosed antenatal. Typical findings on fetal sonogram include bilateral hydroureteronephrosis, a distended bladder, and a dilated prostatic urethra, termed a "keyhole sign".[24] Discrete focal cysts in the renal parenchyma are diagnostic of renal dysplasia. Amniotic fluid volume is variable, and those with normal or slightly reduced amniotic fluid volume have a better prognosis. In contrast, oligohydramnios suggests significant obstructive uropathy or renal dysplasia or both, and pulmonary hypoplasia is common. The presumptive diagnosis of posterior urethral valve cannot be confirmed until postpartum radiological studies are performed.

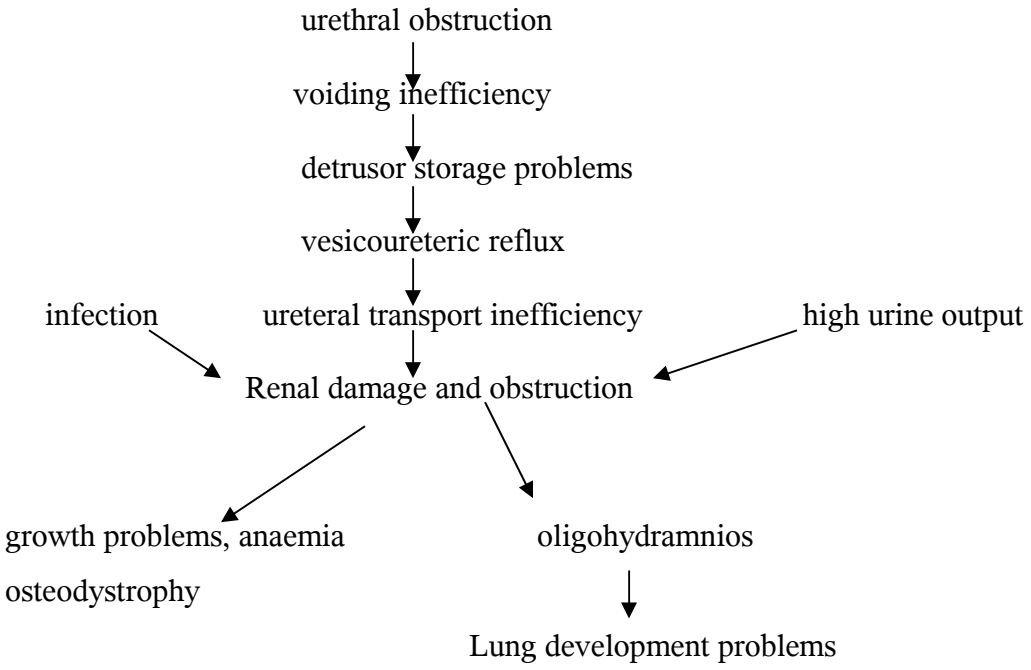
PRIMARY AND SECONDARY PATHOLOGY:

Primary Pathology:

The urethral valves represent a pure obstruction in the urethral conduit; thus, interfering with the voiding efficiency. Urethral obstruction may be complete, so that urinary retention results, or it may be incomplete, so that the energy dissipated in the urethra during urine passage is increased.[25]

Secondary Pathology:

The relatively simple urethral obstruction can lead to a cascading sequence of secondary pathology, beginning with urethral obstruction and ending with inadequate amniotic fluid volume and lung development. It is the level, severity, and reversibility of the secondary pathology that are most important to clinical decisions.[8]



Voiding Efficiency:

Urethral conduit obstruction is not the only pathophysiologic feature interfering with voiding efficiency.[5,8,25] Inadequate detrusor compression, poor coordination of the urethral control mechanism, and other secondary causes of urethral conduit obstruction have been implicated. Urodynamic studies have clearly shown that myogenic failure can result in inefficient voiding.[26] Severe reflux also significantly lowers the efficiency of detrusor compression and conversion of compression energy to kinetic energy of

voiding.[25] Approximately 10 to 15 per cent of patients with posterior urethral valves require supplemental treatment by clean intermittent catheterization to facilitate bladder emptying.[7,8]

Detrusor Storage Capacity:

Clinical reports as well as experimental data show that the single most important determinant of ureteral efficiency is intravesical pressure.[27,28,29] Clinical studies have shown that persistent bladder pressure exceeding 35 cm of water leads to significant ureteral delivery problems and ultimately to renal insufficiency.[25,27,30] Four types of detrusor problems persisting after valve ablation have been identified: small capacity, uninhibited contractions, high pressure voiding, and myogenic failure.[26,31] Valve patients with auxiliary capacity (a pressure "pop-off" mechanism) had much better renal function, as judged by follow-up serum creatinine measurements, than did patients who did not have such a pressure reducing auxiliary capacity. The types of auxiliary capacity in their study included severe unilateral reflux, diverticula, urinary ascites and extravasation.[12]

Vesicoureteric Reflux:

Reflux is detrimental in patients with posterior urethral valves for several reasons:[8]

1. It bypasses the normal bladder infection defense mechanism and can lead to renal scarring and reflux nephropathy.
2. Interferes with detrusor urine compression and conversion of compression energy to kinetic energy so that urine transport from detrusor to atmosphere pressure is less efficient.
3. Causes residual urine[25]

4. Raised ureteral resting pressure and lower ureteral compression pressure[28,29]
5. All above abnormalities may lead to hydronephrosis and irreversible renal damage[30]

Finally, significant reflux is often associated with renal dysplasia and nonfunction.[32] Hoover and Duckett[33], in their study showed that unilateral reflux has a significant left-sided predominance (as high as 75 to 90 per cent).The kidney subjected to reflux is non-functioning in 65 to 90 per cent of cases, and that the contralateral kidney usually has sufficient function even though it is a solitary functioning kidney in 90 per cent of such cases as the kidney affected by reflux acts as a secondary reservoir, a "pop-off" mechanism.

Ureteral Function:

In severe posterior urethral valve, two important mechanical factors- reflux[5] and high pressure detrusor storage[7,8,25], decrease the efficiency of ureteral urine transport. In order to maintain glomerular filtration and renal blood flow, the ureter dilates to maintain a low pressure.[28,29] Ultimately, decompensation occurs because of the lack of ureteral coaption, decreased force generated per unit area, and ureteral tortuosity.

High Urine output:

The urinary tract with inefficient urine transport will have a lower flow for the same pressure gradient as an efficient system and is exaggerated by high urine flow.[25] Polyuria is well recognized in boys with posterior urethral valves.[34] A majority of patients with persistent ureteral dilatation have excessive urinary outputs, approximately two to four times the volume appropriate for their age.

Glassberg, in his study of 15 cases of posterior urethral valves with persistent ureteral dilatation found that all had a concentration defect with an inability to achieve a specific gravity greater than 1.008 after a 14 hour fast. Five of the 15 cases were given a test dose of Pitressin, and all five were unable to achieve urine osmolarities above the serum values, a result that supports a diagnosis of nephrogenic diabetes insipidus.[35]

Jones et al in their study showed that in adults with chronic urinary retention under diuretic conditions, renal pelvic pressures vary with detrusor pressures even in the absence of vesicoureteral reflux, and it is therefore possible that the kidneys of children with high urinary outputs are more susceptible to the effects of high detrusor pressures.[36]

Glassberg et al[37] in their study noted that some upper tracts were obstructed during bladder filling but drained when the bladder was empty. They then attributed the persistent dilatation to a noncompliant, thick-walled bladder and large urine output.

Williams pointed out that some boys with posterior urethral valve have full bladder most of the day and do not sense these fullness. In these boys the full bladder prevents the upper tracts from emptying. The large volume of urine that these boys produce compounds the problem. After the bladder empties these upper tracts are free to drain, and the bladder refills and again obstructs the upper tracts.[38]

Koff et al[39] in their study showed that the cause of valve bladder syndrome is sustained bladder over distension due to a combination of polyuria with high 24-hour urine volume, impaired bladder sensation and high residual urine volume.

Infection:

Hodson and Edwards in their study showed that reflux nephropathy could be produced with urethral obstruction and sterile reflux.[40] However, Hinman found that infection greatly accelerates the renal damage of hydronephrosis.[41] Urinary tract infection, particularly when accompanied by musculomechanical inefficiency, is a significant element for reflux nephropathy to occur but renal damage could also be produced with urethral obstruction and sterile reflux.

Renal Damage and Destruction:

In posterior urethral valves, kidneys are basically damaged by four mechanisms:

I. Hydronephrosis associated with stasis[41]

II. Reflux nephropathy[40]

III. Renal dysplasia[32]

IV. Hyperfiltration and glomerulosclerosis[42]

Oligohydramnios:

When the urinary system is severely obstructed or there is bilateral renal agenesis, the volume of amniotic fluid is markedly decreased resulting in oligohydramnios as 90 per cent of the amniotic fluid is derived from fetal urine.[14]

Lung Development:

Clinical and experimental data suggest that pulmonary hypoplasia is the result of both mechanical factors and chemical factors elaborated by the fetal kidney which causes defective early airway branching and ineffective alveolar development.[14]

Pathophysiology of renal damage in Posterior urethral valves:

There are two pathophysiologic relations, one between the valves and ureteral malfunction, and the other between obstruction and renal dysfunction, which are

responsible for renal damage. The valvular obstruction in the posterior urethra leads to high bladder pressures and progressive bladder thickening, trabeculations, and fibrosis. This bladder dysfunction and maldevelopment lead to distortion of the uretero-vesical junction, causing obstruction, vesicoureteral reflux, or both.[43]

Two types of renal dysfunction are found in valve patients. The more severe type is renal dysplasia, in which the kidney is found to have primitive tubular and glomerular structures, developmental disorganization, and fibrosis.[44] There is decreased, if any, renal function, and the lesion is not treatable or reversible. Renal dysplasia with insufficient function is the most important cause of long-term morbidity and of death in patients with posterior urethral valves. The other form of renal malfunction is less severe and relates to increased pressure in the collecting system and, if the collecting system pressures are relieved early enough, renal function will improve significantly. There is animal experimental evidence that renal dysplasia results from urinary tract obstruction very early in fetal life whereas obstructive uropathy without dysplasia seems to occur when obstruction appears late in gestation.[45]

The late onset of renal failure has been initially thought to be due to the metabolic demands of puberty, but this explanation seems unlikely, as renal deterioration precedes the period of maximal somatic growth.[46] Another possible mechanism for the late onset renal failure is the concept of nephron hyperfiltration. When functional renal mass is reduced, such as in the presence of renal dysplasia, there is compensatory increase in the glomerular filtration rate in the remaining nephrons and an increase in renal size which has been shown experimentally[47] and clinically.[48] Maintenance of whole kidney filtration rates in the presence of continuing renal damage can occur only as a result of increased

single nephron filtration rates (hyperfiltration), as there is no increase in nephron number.[49,50] Studies of rat model have shown that when there is progressive loss of functioning nephrons, eventually a critical point is reached, beyond which any compensatory increase in the single nephron glomerular filtration rate falls further behind the loss in total glomerular filtration and the whole kidney glomerular filtration rate.[51,52]

The changes in Bladder occurring secondary to obstruction:

Bladder dysfunction persists in 75% of boys with posterior urethral valves and it is probably the major determinant of the poor long term prognosis for renal function.[53,54]

Foetal Bladder:

The human bladder is formed by 21 weeks of gestation but for it to mature into an effective filling and emptying organ with the appropriate ratio of collagen type III to type I fibers, some degree of storage is required.[55] Normal mammalian bladder maturation in the latter half of gestation is associated with a decrease in total collagen and in the ratio of collagen type III to type I fibres, associated increase in smooth muscle proliferation, followed by decreased smooth muscle tension, and increase in compliance and storage ability.[56,57] Initially in uterus response to obstruction leads to increase in bladder compliance and contractility. However with more prolonged obstruction the bladder will decompensate and compliance and contractility will fall.[58] At 21 weeks of gestation, just as the lack of distension or storage can be harmful to the bladder development and maturation, so too can over distension or obstruction. It logically seems that the closer to 21 weeks, outlet obstruction manifests the greater the likelihood that normal maturation processes will be compromised, therefore fetuses with a posterior urethral valve diagnosed with hydronephrosis after 24 weeks of gestation have a much

better prognosis than those diagnosed with hydronephrosis before 24 weeks of gestation.[59]

The Obstructed Bladder after birth:

The congenital infravesical obstruction has significant long standing or even permanent deleterious effect on the bladder which can affect the final outcome to a significant extent.[60,61] Valve bladders are thick walled, less elastic, also have poor compliance and may reach higher resting pressures at smaller volumes than normal bladders. Many of these children have high urinary flow rate and may attain unacceptable resting bladder pressures within a short time after voiding. The high bladder pressure impedes ureteral drainage and may itself be the primary factor for persisting ureteral dilatation, rather than uretero-vesical junction obstruction.

Glassberg reported persistent upper urinary tract dilatation following urethral valve ablation, which is most often secondary to a noncompliant thick walled bladder. Persistent incontinence occurs in most of the patients with persistent upper tract dilatation that is secondary to decreased bladder compliance.[62]

Bauer et al reported their findings in 8 boys with a history of posterior urethral valves who had abnormal voiding and found that only 1 had a normal urodynamic study. They divided the bladder findings into myogenic failure, high voiding pressure, uninhibited contractions and small capacity bladder.[60]

Urodynamics in children with Posterior urethral valve:

Voiding dysfunction may occur after valve ablation in 13 to 38% of patients and may not be reversible after relief of obstruction. On urodynamic evaluation of cases with

posterior urethral valve, Beurs and Peters et al identified three primary groups of bladder dysfunction, including

1) myogenic failure, 2) hypertonic, poorly compliant bladders and 3) detrusor instability.[60]

Parkhouse and Woodhouse categorized abnormal urodynamics findings similarly but with slightly different terminology as acontractile bladder, hypocompliance and detrusor instability. A basic pressure increase greater than 20 cm of water on cystometry represents a poorly compliant bladder. Urodynamic abnormalities are present in 20 to 88% of these boys despite adequate relief of urethral obstruction, therefore it is important to have frequent urodynamics follow up, since many of these bladders do not improve on their own and must be managed closely.[63]

Mitchell[64] suggested the term "valve bladder syndrome" to denote association of a noncompliant bladder and upper tract dilatation in boys with a history of posterior urethral valves. Upper tract dilatation most often is secondary to noncompliant thick walled bladder. Incontinence occurs almost always in all the patients and is secondary to decreased bladder compliance and unusual large urine output which is secondary to acquired nephrogenic diabetes insipidus contributing to the upper tract dilatation. As noncompliant bladder is part of the valve bladder therefore regular monitoring with the CMG study is important to decide the course of treatment.

Late Problems:

I. End-stage Renal Disease:

Unfortunately, end-stage renal disease is still a reality for too many patients who survive infancy[5,21,53] The percentage of patients who are in or who will eventually have

end-stage renal disease remain unclear and the longer the patient is followed, the higher the percentage of renal failure becomes.

Parkhouse et al[53] in their study showed that early outcome was statistically as good in patients who had proximal diversion as those who had valve ablation despite the fact that "there was a clinical impression of greater severity of illness in boys who underwent temporary diversion" but had a poorer long term outcome.

Walker et al[65] reported excellent short-term results with vesicostomy (prevesicostomy serum creatinine of 1.8 +/- 1.1 mg/dl and postvesicostomy creatinine of 0.5 +/- 0.4 mg/dl) but long term follow up showed an incidence of renal failure of 21 per cent in childhood.

II. Inadequate Growth

Duckett and Norris stated that the monitoring of growth in patients with posterior urethral valves is mandatory. If the serum creatinine falls below 1 mg/dl during the initial hospitalization and after valve ablation, the prognosis for growth and normal serum creatinine remains good; and that if the postoperative creatinine concentration remains above 1 mg/dl, the likelihood of chronic renal insufficiency and poor growth exists.[6]

III. Hypertension and Proteinuria; possible Hyperfiltration

Hypertension and proteinuria are major prognostic factors in long-term renal outcome. The development of proteinuria or hypertension, especially in a patient whose creatinine clearance is below 25 ml/min/1.73 m^2, is a definite indication that the patient will eventually require dialysis or transplantation.[66]

IV. Urinary Incontinence

Urodynamic studies show that urinary incontinence occurs as a result of increased residual urine volume, small capacity bladder, decreased detrusor compliance, or

abnormal contractility. Furthermore, it showed that if incontinent leakage occurs at a pressure below 35 cm of water then damage to the upper tracts is highly unlikely.[20] Leakage above that pressure will subject the upper tract to the same pressure and probability of inefficient urine transportation and renal damage.[8,27]

V. Polyuria

Polyuria is well recognized in boys with posterior urethral valves.[34] A majority of patients with persistent ureteral dilatation have excessive urinary outputs, approximately two to four times the volume appropriate for their age. Glassberg, in his study of 15 cases of posterior urethral valves with persistent ureteral dilatation found that all had a concentration defect with an inability to achieve a specific gravity greater than 1.008 after a 14 hour fast. Five of the 15 cases were given a test dose of Pitressin, and all five were unable to achieve urine osmolarities above the serum values, a result that supports a diagnosis of nephrogenic diabetes insipidus.[35] Jones et al in their study showed that in adults with chronic urinary retention under diuretic conditions, renal pelvic pressures vary with detrusor pressures even in the absence of vesicoureteral reflux, and it is therefore possible that the kidneys of children with high urinary outputs are more susceptible to the effects of high detrusor pressures.[36]

Reversibility of Secondary Changes:

Bauer and associates, by urodynamics evaluation, have identified three patterns of late bladder dysfunction in patients with posterior urethral valves:[26,31]

1) Myogenic failure with overflow incontinence and large residual volumes;
2) Hyper-reflexic unstable bladder with normal capacity;
3) Small bladder capacity with markedly reduced compliance.

These patients frequently require late treatment consisting of clean intermittent catheterization, pharmacological therapy, and bladder augmentation. Churchill et al[8] have shown that 15 to 20 per cent of patients with posterior urethral valves have such secondary bladder pathology that is not reversible by any type of primary therapy.

Churchill et al[5] in their study found that 40 per cent of the renal units in patients with posterior urethral valves reflux at the time of presentation and was higher in children who presented in the first 30 days (52%) than those presented after 1 year (31%). Among these, 29% of renal units stopped refluxing spontaneously with decompression procedures that did not directly involve the uretero-vesical junction and was independent of the type of decompression used.

The Valve Bladder Syndrome

Mitchell suggested the term "valve bladder syndrome" to denote association of a noncompliant bladder and upper tract dilatation in boys with a history of a posterior urethral valves.[67]

Glassberg et al found that boys with persistent hydronephrosis also had large urine outputs secondary to acquired diabetes insipidus and they were also incontinent secondary to the combination of a large urine output and compliance loss.[38,68] Parkhouse et al found that 85 % of these cases have abnormal bladder dynamics.[53] Persistent hydronephrosis occurred in 82% of those with a component of bladder hypertonia, 40% of those with hyperreflexia and 14% of those with myogenic failure.[69] Kim et al found that patients with decreased bladder compliance responded markedly well to anticholinergic therapy.[70] In the absence of an adequate response to medical therapy a CIC (clean intermittent catheterization) regimen should be used before considering

bladder augmentation. In such cases hypocompliance may improve and become more responsive to anticholinergic therapy. Holmdahl et al advised caution when considering bladder augmentation, because urodynamics pattern may change with time and they found a tendency for bladder hypocompliance to decrease and capacity to increase with age.[71]

Pathophysiologic changes in valve bladder syndrome:[72]

Organ	Pathology	Clinical Effects
Kidney	Dysplasia, renal tubular dysfunction Urine concentrating defect Renal tubular acidosis	Poor renal function; polyuria Rapid filling of bladder, causing persistent hydroureteronephrosis and incontinence Impaired somatic growth, bone demineralization
Ureters	Dilated with poor peristalsis Fibrosis secondary to previous surgery or infection	Large dead space, Increased risk of UTI Poor drainage of upper tract Possible obstruction after ureteric reimplantation
Bladder	Poor compliance, small volume Reduced sensation to high pressure Myogenic failure	High bladder pressure Progressive renal functional damage Incontinence Progressive renal and bladder damage
Bladder neck	Hypertrophy	Poor bladder emptying Voiding dysfunction, Incontinence

Role of Clean intermittent catheterization (CIC) & Overnight Bladder drainage:

Williams pointed out that some boys with posterior urethral valve have full bladder most of the day and do not sense these fullness. In these boys the full bladder prevents the upper tracts from emptying. The large volumes of urine that these boys produce compound the problem. After the bladder empties these upper tracts are free to drain, and

the bladder refills and again obstructs the upper tracts.[38] Post void catheterization or double voiding helps in draining this post void residual urine which is actually pseudo-residual urine representing upper tract drainage.

Koff et al[39] in their study showed that the cause of valve bladder syndrome is sustained bladder over distension due to a combination of polyuria with high 24-hour urine volume, impaired bladder sensation and high residual urine volume. Treatment of over distension during daytime alone was unsuccessful. Nocturnal bladder emptying with an indwelling catheter, intermittent catheterization and/or frequent double voiding markedly improved hydronephrosis, which was comparable to the results after urinary diversion.

Montane et al[73] found that in patients with dysfunctional bladder and progressive polyuric renal failure, continuous overnight bladder drainage delayed progression of renal failure and reduced hospitalization for febrile urinary tract infections.

Anticholinergic drugs:

A major portion of the neurohumoral stimulus for physiologic bladder contraction is acetylcholine induced stimulation of postganglionic parasympathetic cholinergic receptor sites on bladder smooth muscle. Oxybutynin remains the reference standard for anticholinergic medications used to treat incontinence. It has been used effectively in children for many years. It has an efficacy of more than 90% in some series and improves reflux resolution in patients with unstable bladders. In children with neurogenic bladder dysfunction, oxybutynin is effective both in vivo and in vitro.[74] Oxybutynin hydrochloride is administered in a dose of 1.0 mg per year of age every 12 hours to help lower detrusor filling pressures. In neonate and children less than 1 year of age, the dose is lowered to less than 1.0 mg in relation to child's age at the time and increased

proportionately as the age approaches 1 year. Side effects have not been manifest when oxybutynin is administered according to this schedule. All 3 formulation of oxybutynin (tablets, syrup and extended release tablets) are safe and effective in children with neurogenic bladder dysfunction. In these cases, as a rule oxybutynin is employed along with the clean intermittent catheterization.

Intermittent Catheterisation:

Per-urethral clean intermittent self-catheterisation remains the most important single advance in the management of neuropathic bladder in children after its introduction by Lapides et al[75] in 1972. Clean intermittent catheterization (CIC) has proved to be the most effective means of attaining a catheter free state in the majority of patients with acute spinal cord lesions. CIC alone or in combination with anticholinergic agents, when detrusor filling pressures are greater than 40 cm of water and voiding pressures reach levels higher than 80-100 cm of water, has resulted in only an 8-10% incidence of urinary tract deterioration. This represents a significant drop in the occurrence of detrimental changes compared with the group of children followed expectantly.[75,76]

The Mitrofanoff Principle:

Introduced by Mitrofanoff et al[77] in 1980, trans-appendicular continent cystostomies have been widely used to achieve dryness in patients with neurogenic sphincter deficiency. The procedure aims to create a catheterisable continent conduit between the abdominal skin and the bladder, with an antireflux system to prevent leakage. The appendix is favored by most surgeons to create this continent conduit.**Prevention of renal deterioration:**

Prevention of renal deterioration:

The etiology of renal failure associated with posterior urethral valves is multifactorial and that dysplasia with nephron hyperfiltration, polyuria as well as reflux nephropathy, are implicated. The symptom of polyuria is an important predictor of renal tubular function, even in the presence of a normal glomerular filtration rate, and it is likely that those kidneys that are already polyuric are those that are most vulnerable to the adverse effects of raised pressures in the urinary tract.

The implications of these findings are that all boys with urinary incontinence after resection of posterior urethral valves should be investigated urodynamically, as the symptom is more likely to be secondary to detrusor instability than to urethral sphincter incompetence and is worthy of treatment. The successful reduction of detrusor pressures in boys with functional bladder abnormalities could benefit both continence and renal function. Medical treatment with anticholinergic drugs, daytime clean intermittent catheterization with nighttime continuous bladder drainage and surgical treatment with augmentation cystoplasty are good options.

Therefore, early identification of renal tubular defects and urodynamics abnormalities is desirable if any progress is to be made in improving the renal prognosis and quality of life for boys with posterior urethral valves. Urodynamics studies should be done during infancy, and in childhood even in boys who are continent, in order to identify occult detrusor instability at a stage when it can be treated, prior to the onset of renal failure.[40]

MATERIAL AND METHODS

Study Design:

Descriptive study with prospective as well as retrospective limbs.

Study Group:

All children with fulgurated posterior urethral valves and deteriorating renal function undergoing overnight bladder drainage at the Department of Paediatric Surgery, Christian Medical College and Hospital, Vellore between July 2003 and April 2006 were included in this study. From March 2004 onwards the cases were followed prospectively. Children on overnight bladder drainage prior to March 2004 were studied retrospectively with the help of hospital records.

Patient inclusion criteria:

Children with posterior urethral valves after fulguration, who have-

- High 24 hours urinary volumes
- Urinary dribbling
- High post void residual urine
- Persistent upper tract dilatation
- Pronounced bladder dysfunction
- Impaired renal function
- Recurrent urinary tract infections

- With or without rising Serum Creatinine levels

Patients:

During the period of July 2003 and April 2006, overnight bladder drainage program was initiated in 24 boys aged 1 month to 14 years (mean age 5.3 yrs). At the time of initiating overnight bladder drainage, 13 children were on oral anticholinergic (Oxybutynin).

All parents were counseled beforehand regarding the need for clean intermittent catheterization (CIC) and overnight bladder drainage via an indwelling catheter per urethra or Mitrofanoff conduit if one was available. If repeated urethral catheterization was difficult, a Mitrofanoff procedure was done for catheterization. The child or parents were taught to place the catheter into the urethra or Mitrofanoff conduit if one was available overnight from 8 pm to 6 am. At bed time catheter was placed and secured with thread and tape to the penis and inguinal regions. Additionally, the catheters were further secured with tape to lower abdominal wall and connected to urosac for dependent drainage. The child resumed daytime intermittent catheterization the following morning after removal of the catheter.

Patients were evaluated before starting overnight bladder drainage program under following parameters:

1. Age (24 children)
2. Weight (24 children)
3. Height (24 children)
4. Clinical presentation (24 children)
5. Serum Creatinine (24 children)

6. Serum electrolytes (21 children)
7. Serum Calcium/Phosphorus/Alkaline phosphatase (16 children)
8. Urine culture (24 children)
9. Renal sonogram to look for hydroureteronephrosis (24 children)
10. Micturating cystourethrogram to look for vesicoureteric reflux (24 children)
11. Estimation of post void residual urine by ultrasound (in 24 children)
12. Renal isotope scan to look at differential function of kidneys (09 children)
13. Cystometrograms to look at bladder compliance and detrusor instability (12 children)
14. Primary treatment received (24 children)
15. 24 hours urine output (in 23 children)

Patients Characteristics:

1.Age Distribution:

At 1st clinical presentation			At the time of starting OBD		
Age (in months)	Number of cases	Percentage	Age (in years)	Number of cases	Percentage
< 1	4	16.7	< 1	3	12.5
1 to 3	6	25	1 to 2	4	16.6
4 to 6	7	29.1	>2 to 5	8	33.4
7 to 12	3	12.5	>5 to 10	6	25
> 12	4	16.7	> 10	3	12.5
Total	24	100	Total	24	100

Among the 24 cases under present study, only 4 (16.7%) cases presented before 1 month of age. Most of the cases (54%) presented between 1 to 6 months of age with the oldest child being 3 year 11 months. Only 2 children presented during early neonatal period, one at 2nd day of birth and other on 5th day.

In our study, at the time of initiating overnight bladder drainage most of the cases (62.5%) were between 2 to 10 years age group, with 16.6 % cases aged more than 10 years and infants accounted for 12.5 % cases.

2.Geographic Distribution of patient Population:

GEOGRAPHIC LOCATION	NUMBER	PERCENTAGE
Tamilnadu	**11**	**45.8**
Distant States	**10**	**41.7**
Outside India	**3**	**12.5**
TOTAL	**24**	**100**

More than 50% of our cases were from distant places. These patients presented quite late for initial treatment and were relatively more symptomatic. These patients were treated more aggressively and upper tract reconstructive surgery was done at same sitting if necessary as frequent follow up was not possible in them.

3.Socio-economic Status (SES):

Socio-economic Status	NUMBER	PERCENTAGE
LOW	**12**	**50**
MIDDLE	**9**	**37.5**
HIGH	**3**	**12.5**
TOTAL	**24**	**100**

Among all the cases under study 50 % were from low socioeconomic group, 37.5 % from middle and only 12.5 % belonging to high socioeconomic group.

4.Growth Chart: Showing number of cases in relation to weight and height as per Growth chart

	At First presentation		At the time of initiating overnight bladder drainage	
	Weight (kg)	Height (cm)	Weight (kg)	Height (cm)
< 3rd percentile	00	00	02	02
3rd to 50th percentile	13	13	13	13
50th to 97th percentile	11	11	09	09

At first presentation, 13 cases had their weight and height between 3rd to 50th percentiles while 11 children in between 50th to 97th percentile. None of the cases had severe growth retardation or low birth weight.

Among the 24 cases under study 2 children had severe growth retardation with weight and height below the 3rd percentile for that age group, 13 had between 3rd to 50th percentile while 9 children had between 50th to 97th percentile at the time of starting overnight bladder drainage.

5. Clinical Presentation:

Clinical Presentation	**At 1st visit**	**At time of initiating OBD**
Poor urine stream only	**05**	**-**
Poor urine stream, Abd. distension	**02**	**-**
Urinary dribbling only	**-**	**08**
Poor urine stream, Vomiting	**01**	**-**
Abd. distension, Low urine output	**01**	**-**
Poor urine stream, Fever, Convulsion	**01**	**-**
Bed wetting	**-**	**01**
H/o UTI	**-**	**04**
Flank Pain	**-**	**01**
Poor Urine stream, Loose motion	**01**	**-**

Poor Urine stream, Fever, urin.dribbling	**01**	**-**
On Urinary diversion	**01**	**04**
Urin.dribbling, poor stream urine	**01**	**-**
Poor Urine stream, UTI	**09**	**01**
Urinary dribbling, UTI	**01**	**05**
Total	**24**	**24**

Most common clinical presentation in our series prior to starting OBD was urinary dribbling which accounted for 13 cases, with associated UTI in 5 children. History of febrile urinary tract infection was present in 10 cases with one additional child who had positive urine culture without symptoms. Among these 11 cases, 5 presented with urinary dribbling and fever, 4 cases had only fever, 1 case with right side flank pain and pyuria, 1 case with bed-wetting.

6. Serum Creatinine:

	At 1st presentation		At time of starting OBD	
Serum Creatinine (mg/dl)	No. of cases	Percentage	No. of cases	Percentage
upto 0.5	0	0	5	20.8
0.6 to 1.0	7	29.1	12	50
1.1 to 2.0	11	45.9	6	25
>2.0	6	25	1	4.2
Total	24	100	24	100

In our study all cases at initial presentation had raised serum creatinine level, ranging from 0.6 mg/dl to 8.6 mg/dl with mean serum creatinine of 1.87 mg/dl. Of these, 17 (71

%) cases had serum creatinine of more than 1 mg/dl and 7 (29%) cases between 0.6 to 1 mg/dl. At the time of starting OBD, 19 (79%) cases still had serum creatinine more than 0.6 mg/dl and among these 7 cases had more than 1 mg/dl with highest value of 2.9 mg/dl.

7.Co-Morbidities: (n = 24) At time of starting overnight bladder drainage

COMORBIDITY	NUMBER	NO. INVESTIGATED
Hydroureteronephrosis	**24**	**24**
Vesico-ureteric reflux	**12**	**24**
Chronic Renal Failure	**07**	**24**
Poorly/Nonfunc. kidney	**03**	**09**
Poor compliance bladder	**07**	**14**
Significant post void residue	**16**	**24**
Urinary Dribbling	**13**	**24**
H/O UTI	**11**	**24**
On Ureterostomy	**03**	**24**
Poor Growth	**10**	**24**

Most common clinical presentation in our series prior to starting OBD was urinary dribbling which accounted for 13 cases with associated UTI in 5 children.

History of febrile urinary tract infection was present in 10 cases with additional 1 child had positive urine culture. Among these 11 cases, 5 presented with urinary dribbling and fever, 4 cases had only fever, 1 case with right side flank pain and pyuria, 1 case with bed-wetting.

8. Hydroureteronephrosis (HUN):

HUN	At 1st presentation	At time of starting OBD
Bilateral HUN	24	22
Unilateral HUN	00	02
Both kidneys normal	00	00

8.**a) Degree of Hydroureteronephrosis:**

	Right HUN			Left HUN			Total no.of abnormal renal units		
Degree of HUN	**Mild**	**Mod.**	**Severe**	**Mild**	**Mod.**	**Severe**	**Mild**	**Mod.**	**Severe**
At 1st presentation	04	04	16	01	05	18	05	09	34
At time of starting OBD	03	08	12	04	07	12	07	14	25

In our study at time of initiating OBD, all 24 cases had persistent hydroureteronephrosis with bilateral involvement in 22 cases and 2 cases had unilateral mild to moderate HUN. Of these 22 cases with B/L HUN, 11 had severe HUN, 7 had moderate, 2 had mild grade HUN bilaterally and 2 cases had severe HUN on one side and mild HUN on the other side.

9. Vesico Ureteric Reflux (VUR

	At 1st presentation			At time of starting OBD		
Grade of VUR	Unilat.		Bilat.	Unilat.		Bilat.
	Rt.	Lt.		Rt.	Lt.	
1	--	--	--	--	01	
2	--	--	--	--	--	--
3	--	--	--	--	01	--
4	01	--	01	01	--	--
5	02	06	04	02	01	04
VUR to ureterost.level/uret.stump				01	02	01

Among the 24 cases under our study at initial presentation more than half (54%) cases had VUR with 16 renal units having grade 5 VUR and 3 renal units had grade 4 VUR.

In these cases Cystouretherogram done prior to starting OBD showed 11 renal units with grade 5 VUR, in 5 renal units VUR upto ureterostomy site, and 1 renal unit each with grade 1, 3 and grade 4 VUR respectively.

10. Cystometrogram (CMG): 12 cases

The following tables show the bladder compliance and detrusor instability status in pre-OBD stage in 12 cases-

10a).

Bladder Compliance	At the time of initiating OBD
Good	03
Moderate	01
Poor	07
Myogenic failure	01

10b).

Detrusor instability	At the time of initiating OBD
With Detrusor instability	03
Without Detrusor instability	09

Cystometrogram was done in 12 cases prior to initiating OBD. Among these, 7 (58%) cases showed poor compliance bladder, 3 cases had good compliance, 1 case with moderate compliance, and 1 case had large capacity bladder with myogenic failure. 3 cases had associated detrusor instability.

11. Urine output: 23 Cases

24-hrs. Urine output (in ml/kg/hr.)	No. of Cases	Overnight-output (in ml/kg/hr.)	No. of Cases
2 to 4	**15**	**2 to 4**	**10**
4 to 6	**08**	**4 to 6**	**09**
>6	**00**	**>6**	**04**

All cases were having 24 hours urine output above normal for that age group with 13 (59%) children having overnight urine output more than 4 ml/kg/hr.

12. **Post-void Residual Urine:**

% of Total Bladder capacity for age	At the time of initiating OBD
< 10	08
10 to 20	04
>20 (Significant)	12
Total	24

At the time of initiating OBD, 12 (50%) cases had significant volume of post-void residual urine (defined as > 20 % of bladder capacity[39]).

13. **Treatment:**

13a) Primary Treatment

Treatment given	No. of Cases
Fulguration of PUV	13
Bilat.Ureterostomy	4
Fulguration of PUV + Ureterostomy	2
Vesicostomy	2
Fulguration of PUV + Circumcision	1
SPC	1
PUV fulg.+Transuretroureteric anastomosis (TUU) + Uret. Mitrofanoff	1
Total	24

13b) Subsequent Surgeries done:

Surgery done	No. of Cases
Fulguration of PUV	2
Bilat.Ureterostomy	1
Fulguration of PUV + Ureterostomy	1
Unilat. Ureterostomy	1
Bilat.Uret.reimplant + App. Mitrof.	1
Vesicostomy Closure	1
Ureterostomy Closure	1
Ureteric Reimplant	1
PUV Fulguration + Uret.closure	4
TUU + Uret. Mitrofanoff	4
TUU+ Uret.reimplant + Uret. Mitrofanoff	2
Appendicular Mitrofanoff	3
TUU+ Uret.reimplant + App. Mitrofanoff	1
Rt. Nephrectomy+App. Mitrofanoff	1

Primary fulguration of posterior urethral valve was done in 17 (71%) cases, along with ureterostomy on refluxing side ureter in 2 cases, with circumcision in 1 case and in 1 case along with upper tract reconstructive surgery. B/L ureterostomy was done in 4 patients as they presented with urosepsis, high serum creatinine, B/L gross HUN and 3 cases had bilateral major VUR. 1 case who presented with urosepsis, high serum creatinine (3.5 mg/dl), B/L HUN and major VUR was initially treated with suprapubic catheterization, on which child improved and PUV fulguration with loop ureterostomy on the more refluxing side was done on same admission.

2 cases that had grossly elevated serum creatinine with bilateral severe HUN without VUR underwent vesicostomy at initial presentation and later on when they improved PUV fulguration was done. Later on 1 case needed vesicostomy closure while in the other it closed off spontaneously. One child who presented with urosepsis, high serum

creatinine (1.1 mg/dl), B/L moderate HUN, and left side grade 5 VUR with small bladder underwent PUV fulguration along with upper tract reconstructive surgery comprising left to right transuretroureteric anastomosis (TUU) with left lower ureteric stump as Mitrofanoff conduit in same sitting.

Among the cases that had B/L ureterostomy at initial presentation later on all had PUV fulguration along with ureterostomy closure on one side. Subsequently, 1st case had B/L ureteric reimplant with appendicular Mitrofanoff conduit, 2nd case right to left TUU with left ureteric reimplant and appendicular Mitrofanoff conduit, 3rd case had appendicular Mitrofanoff conduit alone while 4th case was still on right ureterostomy.

2 cases who had PUV fulguration as primary treatment presented on follow up with urosepsis along with progressively rising serum creatinine, ureterostomy was done in both cases, bilateral in 1case. Subsequently, 1case had right to left TUU with left ureteric reimplant and right ureteric Mitrofanoff while other case underwent right ureterostomy closure with left nephrectomy.

14. **Conduit Characteristics:**

Conduit for CIC	No. of Cases	Percentage
Urethra	9	37.5
Ureteric Mitrofanoff	9	37.5
Appendicular Mitrofanoff	6	25
Total	24	100.0

Our preferred conduit for CIC in children without major vesicoureteric reflux had been urethra but in those cases where urethral CIC was not possible either due to sensate urethra or difficult urethral catheterization appendicular Mitrofanoff channel was made. In cases with unilateral major vesicoureteric reflux, we used the refluxing lower ureteric stump as Mitrofanoff channel after doing transureteroureteric anastomosis to the other

side nonrefluxing but dilated ureter. In cases with bilateral vesicoureteric reflux, major refluxing side lower ureteric stump was brought out as Mitrofanoff channel and transurteroureteric anastomosis was done to the other side ureter after its reimplantation. In one case that had unilateral major vesicoureteric reflux, we have done Y-ureterostomy with lower stump of the ureter as Mitrofanoff channel that was used for CIC and overnight bladder drainage.

RESULTS

FOLLOW UP PERIOD:

Duration	No. of Cases
< 6 months	2
6 to 12 months	7
1 to 2 years	10
> 2 years	5

All 24 patients were under regular follow up with patients from distant states or outside India, were sending their investigation reports regularly and came for follow up as required.

Mean Follow up = 15 months

Median Follow up = 12 months

Range = 4 months to 3 years

Age distribution:

	At the time of initiating overnight bladder drainage		At last Follow up	
AGE (in years)	NUMBER	PERCENTAGE	NUMBER	PERCENTAGE
< 1	3	12.5	1	4.2
1 to 2	4	16.6	4	16.6
>2 to 5	8	33.4	6	25
>5 to 10	6	25	9	37.6
> 10	3	12.5	4	16.6
TOTAL	24	100	24	100

In our study, at the time of initiating overnight bladder drainage most of the children (62.5%) were between 2 to 10 years age group, with 16.6 % children aged more than 10 years and infants accounted for 12.5 % cases. At the last follow up more than half (54 %) of our children were aged above 5 years with oldest child aged 15 years.

Growth Chart: Showing number of cases in relation to weight and height as per Growth chart

	At the time of initiating overnight bladder drainage		At last Follow up	
	Weight (kg)	Height (cm)	Weight (kg)	Height (cm)
< 3rd percentile	02	02	02	02
3rd to 50th percentile	13	13	12	12
50th to97th percentile	09	09	10	10

Among the 24 cases under study 2 children had severe growth retardation with weight and height below the 3rd percentile for that age group, 12 had between 3rd to 50th percentile while 10 children had between 50th to 97th percentile.

Clinical Presentation:

Clinical Presentation	At time of initiating OBD	At last follow up
Poor Urine stream, UTI	**01**	**00**
Fever	**-**	**01**
Urinary dribbling	**08**	**01**
Bed wetting	**01**	**00**
H/o UTI	**04**	**02**
Flank Pain	**01**	**01**
Low urine output	**-**	**01**
Poor health	**-**	**01**
On Urinary diversion	**04**	**02**
Urinary dribbling, UTI	**05**	**-**
Asymptomatic	**-**	**15**
Total	**24**	**24**

Most common clinical presentation in our series prior to starting OBD was urinary dribbling, which accounted for 13 cases. Among these 13 cases, 5 children had associated UTI.

History of febrile urinary tract infection was present in 10 cases with additional 1 child having only positive urine culture. Among these 11 cases, 5 presented with urinary dribbling and fever, 4 cases had only fever, 1 case with right side flank pain and pyuria, 1 case with bed-wetting. On OBD there was marked improvement in patients with recurrent febrile UTIs. 2 children had 1 episode of febrile UTI while on OBD and required oral antibiotics; both were doing well at last follow up. Only one patient had febrile recurrent UTI while on OBD and had to stop OBD. All cases with recurrent febrile UTI had persistent moderate to severe hydroureteronephrosis but no significant change in serum creatinine level.

At last follow up most of the children were doing well clinically with resolution of urinary dribbling in all except 1 case who has occasional urinary dribbling,

Serum Creatinine:

	At time of initiating OBD		At last Follow up	
Serum Creatinine (mg/dl)	No. of cases	Percentage	No. of cases	Percentage
upto 0.5	5	20.8	6	25
0.6 to 1.0	12	50	13	54.2
1.1 to 2.0	6	25	1	4.2
>2.0	1	4.2	4	16.6
Total	24	100	24	100

At the time of starting OBD, 19 (79%) cases had serum creatinine level of more than 0.6 mg/dl and among these 7 cases had more than 1 mg/dl with highest value of 2.9 mg/dl. At last follow up, 5 cases had serum creatinine level of more than 1 mg/dl and these were the cases who had the serum creatinine level 0.9,1.7,1.8,1.9, and 2.9 mg/dl respectively at the time of initiating OBD. Only 3 cases with serum creatinine level of more than 1 mg/dl prior to OBD showed improvement with level decreasing from 1.7,1.3, and 1.1 mg/dl to 0.8,0.8, and 0.6 mg/dl respectively.

We found that among the 7 cases who had serum creatinine level of more than 1mg/dl prior to initiating OBD, 4 cases deteriorated further and showed progressive rise while 3 cases showed marked improvement in serum creatinine level on OBD. In these 4 cases with rising trend of serum creatinine, OBD was started quite late with age of children

being 5 yr.8 months, 7 yrs., 7 yrs. and 13 yrs. respectively. In most of the cases where OBD was started at relatively younger age, serum creatinine either remained stable or showed improvement. In 2 cases where we had started OBD at the age of 4 years with serum creatinine of 1.7mg/dl and 1.3 mg/dl at that time, after a follow up period of 2 yr.3 months and 1 yr. respectively both children were doing well with serum creatinine stabilized at 0.8 mg/dl.

Co-Morbidities: (n = 24) At last Follow up

COMORBIDITY	NUMBER OF CASES	NO. INVESTIGATED
HUN/HDN	**23**	**24**
Vesicoureteric reflux	**03**	**23**
Chronic Renal Failure	**05**	**24**
Poor compliance bladder	**03**	**14**
Significant post void residue	**07**	**24**
Urinary Dribbling	**01**	**24**
H/O UTI	**03**	**24**
On Ureterostomy	**02**	**24**
Poor Growth	**02**	**24**

At the last follow up most of the children were doing well clinically with resolution of urinary dribbling in all except 1 case who has occasional urinary dribbling. On OBD there was marked improvement in patients with recurrent febrile UTIs. 2 children had 1 episode of febrile UTI while on OBD and required oral antibiotics; both were doing well at last follow up. Only one patient had symptomatic recurrent UTIs while on OBD and had to stop OBD. All cases with recurrent febrile UTIs had persistent moderate to severe hydroureteronephrosis but no significant change in serum creatinine level.

Hydroureteronephrosis (HUN):

No. of Cases

HUN	At time of starting OBD	At last Follow up
Bilateral HUN	22	19
Unilateral HUN	02	04
Both kidneys normal	00	01

Degree of Hydroureteronephrosis:

	Right HUN			Left HUN			Total no. of abnormal renal units		
Degree of HUN	**Mild**	**Mod.**	**Severe**	**Mild**	**Mod.**	**Severe**	**Mild**	**Mod.**	**Severe**
At time of starting OBD	03	08	12	04	07	12	07	15	24
At last Follow up	08	10	04	07	10	05	15	20	09

In our study at time of initiating OBD, all 24 cases had persistent hydroureteronephrosis with bilateral involvement in 22 cases and 2 cases had unilateral mild to moderate HUN. Of these 22 cases with B/L HUN, 11 had severe HUN, 7 had moderate, 2 had mild grade HUN bilaterally and 2 cases had severe HUN on one side and mild HUN on the other side.

At last follow up, among the 11 cases with pre-OBD B/L severe HUN, 13 renal units showed some improvement in degree of hydroureteronephrosis but 9 renal units showed either deterioration or persistent gross HUN. The results were much better in those cases with mild to moderate HUN at pre-OBD stage and all cases showed improvement in grade of HUN with 3 renal units having complete resolution.

Vesicoureteric Reflux (VUR):

	At time of starting OBD			At last Follow up		
	Unilat.		Bilat.	Unilat.		Bilat.
Grade of VUR	Rt.	Lt.		Rt.	Lt.	
1	--	01	--			
2	--	--	--			
3	--	01	--			
4	01	--	--	01		
5	02	01	04			
VURto ureterost.level/uret.stump	01	02	01	01	02	01

Course of Vesicoureteric Reflux:

	No. of Renal units involved					
Grade of VUR	**At 1st presentation**	**At time of starting OBD**	**At last Follow up**	**Required surgery**	**Improved without surgery**	**Still persisting**
1		**01**			**01**	
2						
3		**01**		**01**		
4	**03**	**01**	**01**			**01**
5	**16**	**11**		**11**		
To ureterost. site/uret.stump		**05**	**05**			**05**
Total	**19**	**19**	**06**	**12**	**01**	**06**

Type of surgery done for Vesicoureteric reflux:

Type of surgery	No. of Cases
Ureteric reimplant	**01**
Transureteroureteric anastomosis (TUU)	**04**
TUU + Uret. reimplant	**02**

Among the 24 cases under our study more than half (54%) cases had VUR at their initial clinical presentation with 14 renal units having grade 5 VUR and 3 renal units with grade 4 VUR. Among these, cystourethrogram done prior to starting OBD showed 11 renal units with grade 5 VUR, 5 renal units with VUR upto ureterostomy site, and 1 renal unit each with grade 1, 3 and Grade 4 VUR respectively.

Follow up Cystourethrogram was done in only those 12 cases that had VUR prior to initiating OBD. The study showed VUR in only 6 renal units of which 4 renal units had VUR to the ureterostomy site, 1 to ureteric stump left behind after nephrectomy, and 1 child had right side Grade 4 VUR.

Post-void Residual Urine:

% of Total Bladder capacity for age	At the time of initiating OBD	At last Follow up
< 10	08	13
10 to 20	04	04
>20 (Significant)	12	07
Total	24	24

At the time of initiating OBD, 12 (50%) cases had significant volume of post-void residual urine (defined as > 20 % of bladder ca (D).

At last follow up we recorded reduction in post-void residual urine in 22 (92%) cases, with 1 case showing no significant change while another case had increased amount of post-void residual urine.

Compliance with Overnight Bladder drainage:

The following table shows the figures-

Compliance	Number	Percentage
Good	**22**	**91.6**
Irregular	**01**	**4.2**
Stopped	**01**	**4.2**
Total	**24**	**100**

One child had stopped overnight bladder drainage 2 months back due to recurrent urinary tract infection. This child was on right Y-ureterostomy, which was refluxing, giving adequate bladder decompression with insignificant post void residual urine, decreasing hydroureteronephrosis and normal serum creatinine level.

One child who was irregular with overnight bladder drainage, had difficulty in urethral catheterization whom we readmitted for CIC training and presently child himself is doing CIC quite comfortably.

Cystometrogram (CMG): 12 cases

The following tables show the bladder compliance and detrusor instability status in pre- and post-OBD stages in 12 cases-

Bladder Compliance	**At the time of initiating OBD**	**At last Follow up**
Good	03	06
Moderate	01	02
Poor	07	04
Myogenic failure	01	00

Detrusor instability	**At the time of initiating OBD**	**At last Follow up**
With Detrusor instability	03	02
Without Detrusor instability	09	10

Cystometrogram was done in 12 cases prior to initiating OBD. Among these, 7 (58%) cases showed poor compliance bladder, 3 cases had good compliance, 1 case with moderate compliance, and 1 case had large capacity bladder with myogenic failure. 3 cases had associated detrusor instability.

On follow up Cystometrogram, there was some improvement in bladder compliance with 6 cases having good compliance bladder, 2 cases moderate compliance, and 4 cases had poor compliance bladder. There was associated detrusor instability in 2 cases.

There was improvement in bladder compliance in 4 cases and 1 case showed improvement in detrusor instability.

Relationship between Age at initiating OBD, Time interval from 1st presentation and follow up Serum Creatinine:

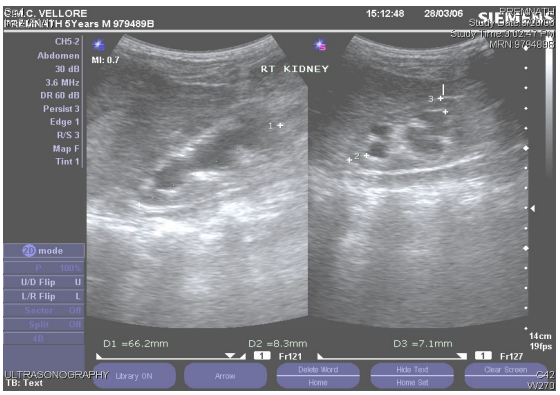
Serum Creatinine (in mg/dl) - on Follow up

Case No.	Age at initiating OBD (in years)	Interval between 1st presentation to starting OBD(in years)	At1st presentation	Pre-OBD period	At starting OBD	Post-OBD period	At last Follow up
1	0.3	0.1	2.7	0.8	0.7	0.6	0.6
2	3	2.9	1.6	0.4	0.6	0.6	0.8
3	1.2	0.8	1.4	0.7	0.7	0.6	0.7
4	14	13.8	1.1	0.3	0.9	0.8	0.9
5	7.5	7.2	1.2	0.6	0.7	0.9	0.6
6	1.6	1.5	8.6	0.9	0.4	0.5	0.4
7	13	10.5	0.8	1.4	1.8	3.3	3
8	7	6.8	1.5	0.8	1.7	2.2	2.1
9	7	6.7	1.3	1.2	1.9	1.2	2.4
10	12	11.9	1.2	0.7	0.9	1.1	0.9
11	4.6	4.1	1.5	2.1	0.8	0.6	0.7
12	8.7	8.2	0.9	0.6	0.7	0.7	0.7
13	10	9	0.7	0.6	0.7	0.6	0.6
14	5	4.9	0.9	0.4	0.5	0.5	0.5
15	4	3.3	2.4	1.3	1.7	0.8	0.8
16	4	0.1	1.3	-	1.3	0.9	0.8
17	3.3	0.9	0.6	-	0.5	-	0.5
18	5.7	5.5	3.8	2.2	2.9	7.1	3.7
19	0.6	0.2	1.1	0.7	0.5	0.5	0.5
20	5	4.5	1.3	0.9	0.9	1.0	1.1
21	2	0.5	0.7	-	0.5	-	0.4
22	1	0.1	0.7	0.6	0.7	-	0.7
23	0.1	0.1	3.5	0.6	1.1	0.6	0.6
24	2.3	2.3	4	0.8	0.7	-	0.5

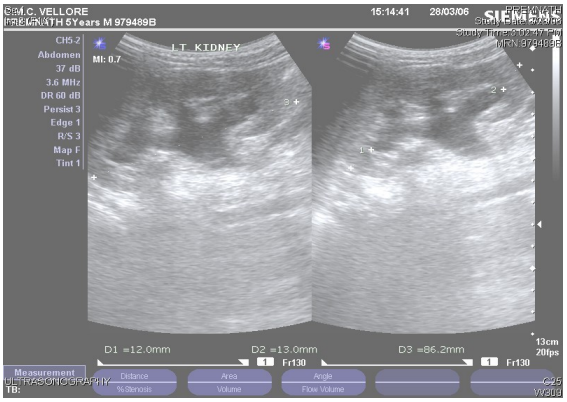
CASE-15 RENAL USG (OBD started at the age of 4 yrs.)

AT PRE-OBD STAGE **AT POST-OBD STAGE**

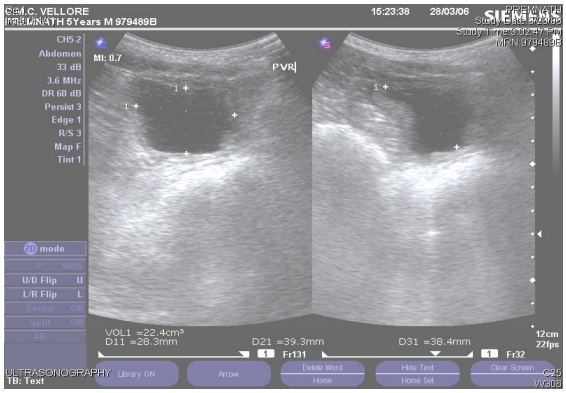
RIGHT KIDNEY



LEFT KIDNEY



URINARY BLADDER

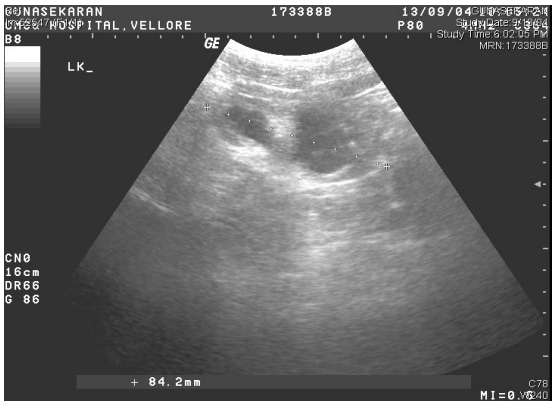


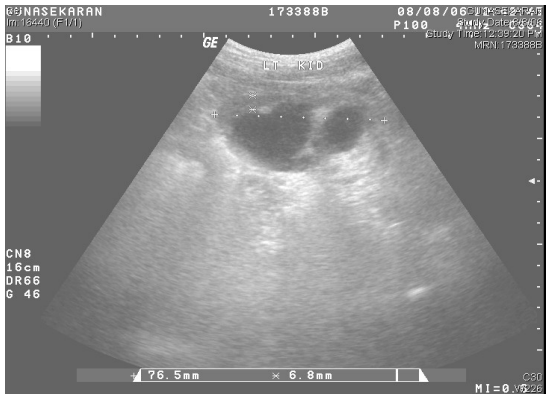
Showing marked reduction in the degree of HUN at Post-OBD stage

CASE-7 RENAL USG (OBD STARTED AT THE AGE OF 13 YRS.)

AT PRE-OBD STAGE **AT POST-OBD STAGE**

RIGHT KIDNEY





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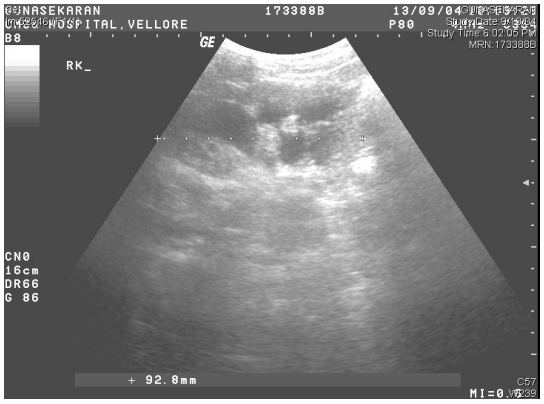

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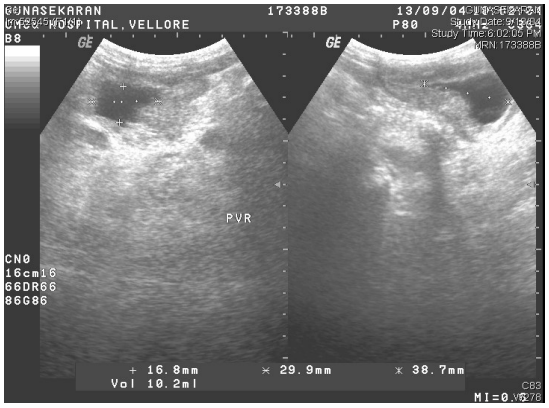
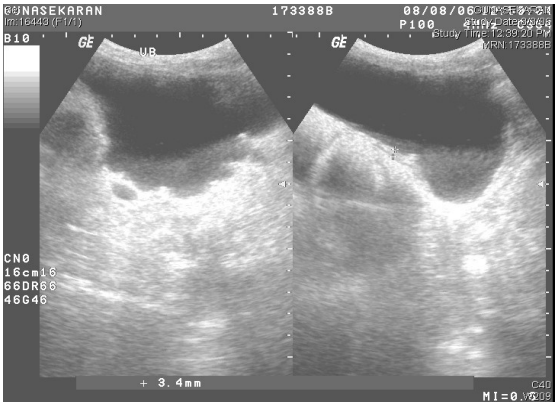




Showing increase in the degree of HUN at Post-OBD stage

Progress of serum Creatinine level in relation to time interval from initial presentation to initiation of OBD:

Among the 11 cases, where the time interval was less than 3 years from initial presentation to initiation of OBD the serum creatinine level had either decreased or remained same. This is in contrast to cases with time interval more than 5 years where among 9 cases none of them showed improvement in serum creatinine level, 4 cases had increased level and in 5 cases serum creatinine level was almost same. In 4 cases where time interval was from 3 to 5 years, 2 cases had almost same level while 1 case each showed increased and decreased level of serum creatinine.

Progress of serum Creatinine level in relation to age of children at the time of initiation of OBD:

Among the 13 cases in the age group less than 5 years, all cases showed either decreased or same level of serum creatinine on post-OBD follow up period. This is in contrast to cases where OBD was started when they were of 5 years age or more. In this age group among the 11 children, 5 showed increased level of serum creatinine while in remaining 6 cases serum creatinine level was almost same.

Current Management:

Current Management	No. of Cases*
Overnight bladder drainage (OBD) only	3
Oxybutynin + OBD	4
OBD+ Daytime CIC	7
Oxyb. + OBD+ Daytime CIC	8
Oxyb. + OBD +Daytime frequent voiding	1

*** 1 child stopped Overnight bladder drainage due to Recurrent UTI**

Among the 24 cases on overnight bladder drainage, prior to starting OBD 8 cases were on oxybutynin with day time CIC, 7 cases on day time CIC, 4 cases on oxybutynin alone and 1 case was on oxybutynin with day time frequent voiding. All the cases except one were doing CIC and OBD regularly as trained. One child has stopped overnight bladder drainage 2 months back due to recurrent urinary tract infection. This child was on right Y-ureterostomy, which was refluxing, giving adequate bladder decompression with insignificant post void residual urine, decreasing hydroureteronephrosis and normal serum creatinine level at the last follow up.

Complications:

Complications	No. of Cases
UTI	03
Difficult Catheterization	01
Urine leak from Mitrofanoff stoma	01

On OBD there was marked improvement in patients with recurrent febrile UTIs. 2 children had 1 episode of febrile UTI while on OBD and required oral antibiotics; both were doing well at last follow up. Only one patient had symptomatic recurrent UTIs while on OBD and had to stop OBD. All cases with febrile UTIs had persistent moderate to severe hydroureteronephrosis without any significant change in serum creatinine level. One child who was irregular with overnight bladder drainage, had difficulty in urethral catheterization whom we readmitted for CIC training and presently child himself is doing CIC quite comfortably.

One child who was on ureteric Mitrofanoff presented with occasional leakage of urine from stoma site, which stopped on its own.

Conduit Characteristics:

Size of Catheter used for CIC :

All except 1 child were using number 8 infant feeding tubes for CIC. One infant was using number 6 infant feeding tubes for urethral CIC**.**

CIC done by:

11 children were performing CIC with ease by themselves. Among these, all children were of 5 years age or more except one child aged 3 ½ years who was doing CIC through ureteric Mitrofanoff quite comfortably.

13 children had one of their parents doing CIC mostly the mother. Only 1 child in this group was older than 5 years (7 years old) but was still too anxious to do CIC himself.

Patient Satisfaction and Social acceptance of the Child:

Social acceptance of the child was assessed by school attendance of children aged more than 5 years. All children above 5 years age were attending school regularly.

Patients and family were overall satisfied with the CIC and overnight bladder drainage.

8 children aged 6 to 15 years who were doing CIC through Mitrofanoff stoma were asked about their views on the CIC and overnight bladder drainage and how it has affected their lifestyle.

The questions asked were:

- Does the child feel satisfied with the operation?
- Does the child perceive any alteration of body image due to Mitrofanoff stoma?
- Is he accepted well at school and among friends?

- Would the child recommend similar treatment options to another child if needed?

RESPONSE TO QUESTION ASKED	NUMBER OF CHILDREN
Satisfied with the operation	8
Perceive some alteration of body image	3
Well accepted at school & among friends	8
Would recommend this surgery to another child	7

All 8 children were satisfied with the operation. They as well as their parents felt that there was definite improvement in their quality of life after starting CIC and overnight bladder drainage. 3 out of 8 children perceived some alteration of body image due to the stoma. All were well accepted at school and among friends; and were playing normally with other children. Only 1 child aged 7 years said he would not recommend this operation to another child, and was still too anxious to do CIC himself (being done by mother).

DISCUSSION

In our study we evaluated the use of overnight bladder drainage (OBD) in 24 children with posterior urethral valves who demonstrated bladder dysfunction with evidence of varying stages of renal compromise. Even after ablation of posterior urethral valves, some patients continue to suffer from increasing hydronephrosis, increase in serum creatinine, recurrent urinary tract infections and urinary incontinence. Major vesicoureteric reflux is present in 50% of the children and there is also late onset of renal function deterioration in about 1/3rd of cases. The underlying problem is bladder dysfunction, termed "Valve Bladder" which is classified into hypo compliant, hyperreflexic and myogenic failure types. However, combinations are common and one type may lead to another. Poor compliance bladder has been associated with the worst prognosis but other factors implicated are age at diagnosis, renal dysplasia, renal function before and after treatment to remove the obstruction, associated vesicoureteric reflux, urinary tract infection, hypertension and initial treatment received.[39,77]

Age Distribution:

In our study, at the time of initiating overnight bladder drainage most of the cases (62.5%) were between 2 to 10 years age group, with 16.6 % cases aged more than 10 years and infants accounted for 12.5 % cases. At the last follow up more than half (54 %) of our cases were aged above 5 years with 1/3rd of cases between 5 to 10 yrs.of age with dribbling of urine being the most common indication for starting OBD. The youngest child in our series was a 1 month old infant whom we put on CIC with OBD as the serum creatinine even after adequate PUV fulguration and left low loop ureterostomy

remained high (1.1 mg/dl) with thick palpable bladder. At last follow up, the child now 8 months old, was doing well with serum creatinine of 0.6 mg/dl.

We are now initiating it in patients at a much younger age, not only to prevent the development of hydroureteronephrosis and the valve bladder syndrome but also to preserve renal function by eliminating bladder over distension.

Growth pattern:

Among the 24 cases under study 2 children had severe growth retardation with weight and height below the 3rd percentile for that age group, 12 had between 3rd to 50th percentile while 10 children had between 50th to 97th percentile. All children with serum creatinine more than 1 mg/dl after primary treatment and at last follow up showed poor growth. We did not find any significant change in the growth pattern in our children as the duration of follow up was short. However, in the long run by preserving renal function it may be possible to improve the growth pattern.

Duckett and Norris stated that the monitoring of growth in patients with posterior urethral valves is mandatory; if the serum creatinine falls below 1 mg/dl during the initial hospitalization and after valve ablation, the prognosis for growth and normal serum creatinine remains good; and that if the postoperative creatinine concentration remains above 1 mg/dl, the likelihood of chronic renal insufficiency and poor growth exists.[6]

Urinary incontinence:

In all 13 patients with urinary incontinence in our study there was complete resolution or improvement of the incontinence on overnight bladder drainage. This was probably because of elimination of the residual urine and improvement in the bladder compliance. Prior to starting overnight bladder drainage urodynamic studies were done in 8 of these

patients, which showed poor compliance bladder in 5 cases, myogenic failure in 1 cases and good compliance bladder in 2 cases.

Urodynamic studies at last follow up in these cases showed some improvement with 4 cases having good compliance bladder, 2 cases moderate compliance bladder and 2 cases with poor compliance bladder. Therefore overnight bladder drainage, by improving bladder compliance, improved the urinary dribbling.

Urodynamic studies show that urinary incontinence occurs as a result of increased residual urine volume, small capacity bladder, decreased detrusor compliance, or abnormal contractility. Furthermore, it showed that if incontinent leakage occurs at a pressure below 35 cm of water then damage to the upper tracts is highly unlikely.[20] Leakage above that pressure will subject the upper tract to the same pressure and probability of inefficient urine transportation and renal damage exists.[8,27]

24 hours Urine output and Overnight Bladder drainage(OBD)output:

In our study we found that all cases had 24 hours urine output above the normal range with one-third cases having more than 4 ml/kg/hr. The overnight bladder drainage was markedly elevated in more than half of our cases with 4 cases having more than 6 ml/kg/hr.

Polyuria is well recognized in boys with posterior urethral valves.[34] A majority of patients with persistent ureteral dilatation have excessive urinary outputs; approximately two to four times the volume appropriate for their age. Glassberg, in his study of 15 cases of posterior urethral valves with persistent ureteral dilatation found that all had a concentration defect with an inability to achieve a specific gravity greater than 1.008 after a 14 hour fast. Five of the 15 cases were given a test dose of Pitressin, and all five were

unable to achieve urine osmolarities above the serum values, a result that supports a diagnosis of nephrogenic diabetes insipidus.[35] Jones et al in their study showed that in adults with chronic urinary retention under diuretic conditions, renal pelvic pressures vary with detrusor pressures even in the absence of vesicoureteral reflux, and it is therefore possible that the kidneys of children with high urinary outputs are more susceptible to the effects of high detrusor pressures.[36]

Koff et al[39] in their study showed that the cause of valve bladder syndrome is sustained bladder over distension due to a combination of polyuria with high 24-hour urine volume, impaired bladder sensation and high residual urine volume.

Post void residual urine:

At the time of initiating OBD, 12 (50%) cases had significant volume of post-void residual urine (defined as > 20 % of bladder capacity).[39] We recorded reduction in post-void residual urine in 22 (92%) cases, with 1 case showing no significant change while another case had increased amount of post-void residual urine.

Williams pointed out that some boys with posterior urethral valve have full bladder most of the day and do not sense these fullness. In these boys the full bladder prevents the upper tracts from emptying. The problem is compounded by the large volume of urine that these boys produce. After the bladder empties these upper tracts are free to drain, and the bladder refills and again obstructs the upper tracts.[38] Post void catheterization or double voiding helps in draining this post void residual urine which is actually pseudo-residual urine representing upper tract drainage.

Koff et al[39] in their study showed that the cause of valve bladder syndrome is sustained bladder over distension due to a combination of polyuria with high 24-hour urine volume,

impaired bladder sensation and high residual urine volume. Treatment of over distension during daytime alone was unsuccessful. Nocturnal bladder emptying with an indwelling catheter, intermittent catheterization and/or frequent double voiding markedly improved hydronephrosis, which was comparable to the results after urinary diversion.

Serum Creatinine:

At the time of starting OBD, 19 (79%) cases had serum creatinine level of more than 0.6 mg/dl and among these 7 cases had more than 1 mg/dl with highest value of 2.9 mg/dl. At last follow up 4 cases had serum creatinine level more than 2 mg/dl and these were the cases who had the serum creatinine level 1.7,1.8,1.9, and 2.9 mg/dl respectively at the time of initiating OBD. Only 3 cases with serum creatinine level of more than 1 mg/dl prior to OBD showed improvement with level decreasing from 1.7,1.3, and 1.1 mg/dl to 0.8,0.8, and 0.6 mg/dl respectively.

Among the 11 cases, where the time interval was less than 3 years from initial presentation to initiation of OBD the serum creatinine level had either decreased or remained same. This is in contrast to cases with time interval more than 5 years where among 9 cases none of them showed improvement in serum creatinine level, 4 cases had increased level and in 5 cases serum creatinine level was almost same. In 4 cases where time interval was from 3 to 5 years, 2 cases had almost same level while 1 case each showed increased and decreased level of serum creatinine.We found that among the 7 cases who had serum creatinine level of > 1mg/dl prior to initiating OBD 4 cases deteriorated further and showed progressive rise while 3 cases showed marked improvement in serum creatinine level on OBD. In these 4 cases with rising trend of serum creatinine, OBD was started quite late with age of children being 5 yr.8 months, 7

yrs., 7 yrs. and 13 yrs. respectively. In most of the cases where OBD was started at relatively younger age, serum creatinine either remained stable or showed improvement. In 2 cases where we had started OBD at the age of 4 years with serum creatinine of 1.7mg/dl and 1.3 mg/dl at that time, after a follow up period of 2 yr.3 months and 1 yr.respectively both children were doing well with serum creatinine stabilized at 0.8 mg/dl. Therefore in order to prevent renal deterioration OBD program should be considered at a much earlier age after the fulgaration of valves.

Duckett and Norris stated if the serum creatinine falls below 1 mg/dl during the initial hospitalization and after valve ablation, the prognosis for growth and normal serum creatinine remains good; and that if the postoperative creatinine concentration remains above 1 mg/dl, the likelihood of chronic renal insufficiency and poor growth exists.[6]

Montane et al[72] found that in patients with dysfunctional bladder and progressive polyuric renal failure, continuous overnight bladder drainage delayed progression of renal failure.

Hydroureteronephrosis:

In our study at the time of initiating OBD, all 24 cases had persistent hydroureteronephrosis with bilateral involvement in 22 cases and 2 cases had unilateral mild to moderate HUN. Of these 22 cases with B/L HUN, 11 had severe HUN, 7 had moderate, 2 had mild grade HUN bilaterally and 2 cases had severe HUN on one side and mild HUN on the other side.

At last follow up among the 11 cases with pre-OBD B/L severe HUN, 13 renal units showed some improvement in degree of hydroureteronephrosis but 9 renal units showed either deterioration or persistent gross HUN. The results were much better in those cases with mild to moderate HUN at pre-OBD stage and all cases showed improvement in

grade of HUN with 3 renal units having complete resolution. Therefore it is necessary to start the OBD before the onset of gross hydroureteronephrosis when it becomes irreversible.

Glassberg et al[37] in their study observed a profound change in the degree of hydroureteronephrosis in some boys with a history of previously ablated posterior urethral valves after catheter drainage. They noted that some upper tracts were obstructed during bladder filling but drained when the bladder was empty. They then attributed the persistent dilatation to a noncompliant, thick-walled bladder and large urine output.

Williams pointed out that some boys with posterior urethral valve have full bladder most of the day and do not sense these fullness. In these boys the full bladder prevents the upper tracts from emptying. The problem is compounded by the large volumes of urine that these boys produce. After the bladder empties these upper tracts are free to drain, and the bladder refills and again obstructs the upper tracts.[38] Post void catheterization or double voiding helps in draining this post void residual urine, which is actually pseudo-residual urine representing upper tract drainage.

Vesicoureteric Reflux(VUR):

Among the 24 cases under our study more than half (54 %) cases had VUR at their initial clinical presentation with 16 renal units having grade 5 VUR and 3 renal units with grade 4 VUR. Among these, Cystourethrogram done prior to starting OBD showed 11 renal units with grade 5 VUR, 5 renal units with VUR upto ureterostomy site, and 1 renal unit each with grade 1,3 and 4 VUR respectively.

Follow up Cystourethrogram study showed VUR in only 6 renal units of which 4 renal units had VUR to ureterostomy site, 1 to ureteric stump left behind after nephrectomy,

and 1 child had right side grade 4 VUR. This improvement in VUR was mostly because of surgical correction.

Vesicoureteric Reflux is detrimental in patients with posterior urethral valves for several reasons:[8]

6. It bypasses the normal bladder infection defense mechanism and can lead to renal scarring and reflux nephropathy.
7. Causes residual urine[25]
8. Raised ureteral resting pressure and lower ureteral compression pressure[28,29]
9. All above abnormalities may lead to hydronephrosis and irreversible renal damage[30]

Hoover and Duckett,[33] in their study showed that unilateral reflux has a significant left-sided predominance (as high as 75 to 90 per cent). The kidney subjected to reflux is non-functioning in 65 to 90 per cent of cases, and that the contralateral kidney usually has sufficient function even though it is a solitary functioning kidney in 90 per cent of such cases as the kidney affected by reflux acts as a secondary reservoir, a "pop-off" mechanism.

Cystometric Study:

Cystometrogram was done in 12 cases prior to initiating OBD. Among these, 7 (58%) cases showed poor compliance bladder, 3 cases had good compliance, 1 case with moderate compliance, and 1 case had large capacity bladder with myogenic failure. 3 cases had associated detrusor instability.

On follow up Cystometrogram, there was some improvement in bladder compliance with 6 cases having good compliance bladder, 2 cases moderate compliance, and 4 cases had poor compliance bladder. There was associated detrusor instability in 2 cases.

By initiating overnight bladder drainage, thereby keeping the bladder empty helped in improving the bladder compliance in some of the cases.

On urodynamic evaluation of cases with posterior urethral valve, Beurs and Peters et al[60] identified three primary groups of bladder dysfunction, including

2) Myogenic failure, 2) hyper tonic, poorly compliant bladders and 3) detrusor instability

Parkhouse and Woodhouse[54] categorized abnormal urodynamics findings similarly but with slightly different terminology as acontractile bladder, hypo compliance and detrusor instability. A basic pressure increase greater than 20 cm of water on cystometry represents a poorly compliant bladder. Urodynamic abnormalities are present in 20 to 88% of these boys despite adequate relief of urethral obstruction, therefore it is important to have frequent urodynamics follow up, since many of these bladders do not improve on their own and must be managed closely.

Mitchell[64] suggested the term “valve bladder syndrome” to denote association of a noncompliant bladder and upper tract dilatation in boys with a history of posterior urethral valves. Upper tract dilatation most often is secondary to noncompliant thick walled bladder. Incontinence occurs almost always in all the patient and is secondary to decreased bladder compliance and unusual large urine output which is secondary to acquired nephrogenic diabetes insipidus contributing to the upper tract dilatation. As

noncompliant bladder is part of the valve bladder therefore regular monitoring with the CMG study is important to decide the course of treatment.

Complications:

In our study, 11 cases had history of symptomatic UTIs prior to starting OBD. Among these 11 cases, 5 presented with urinary dribbling and fever, 4 cases had only fever, 1 case with right side flank pain and pyuria, and 1 case with bed-wetting.

On OBD there was marked improvement in patients with recurrent febrile UTIs. 2 children had 1 episode of febrile UTI while on OBD and required oral antibiotics; both were doing well at last follow up.

Only one patient had symptomatic recurrent UTIs while on OBD and had to stop OBD. All cases with recurrent febrile UTIs had persistent moderate to severe hydroureteronephrosis but no significant change in serum creatinine level.

Therefore, the initial fears that repeated catheterization might produce recurrent febrile UTIs was not true. On the other hand OBD resulted in a reduction in the incidence of UTIs probably by eliminating the residual urine.

Montane et al[73] similarly found that in patients with dysfunctional bladder and progressive polyuric renal failure, continuous overnight bladder drainage delayed progression of renal failure and reduced hospitalization for febrile urinary tract infections.

One child who was irregular with overnight bladder drainage, had difficulty in urethral catheterization whom we readmitted for CIC training and presently child himself is doing CIC quite comfortably.

One child who was on ureteric Mitrofanoff presented with occasional leakage of urine from stoma site, which stopped on its own.

Social and Family acceptance:

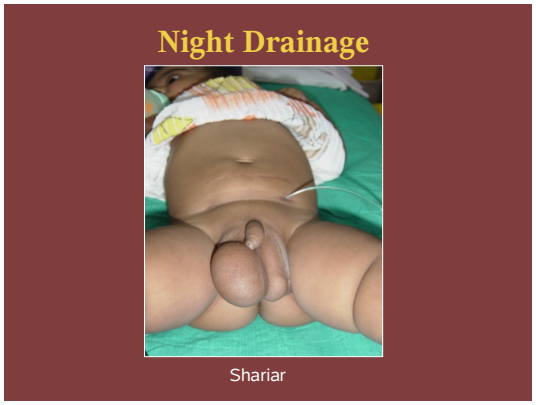
All children above 5 years age were attending school regularly.

They as well as their parents felt that there was definite improvement in their quality of life after starting CIC and overnight bladder drainage. All were well accepted at school and among friends; and were playing normally with other children. Sometimes these children find it difficult to do CIC at schools where proper toilet facilities are not available. Patients and family were overall satisfied with the CIC and overnight bladder drainage.

CHILDREN ON CIC AND OVERNIGHT BLADDER DRAINAGE (OBD)

ON APPENDICULAR MITROFANOFF

ON URETERIC MITROFANOFF



ON URETERIC MITROFANOFF

ON URETHRAL OBD

AT LAST! HAPPY CHILD- SATISFIED MOTHER

CONCLUSION

On the basis of our limited experience we believe that nocturnal bladder emptying is safe and may prove to be a preferred alternative to surgical treatment of boys with the valve bladder syndrome as it specifically treats the pathophysiological mechanisms that cause the syndrome. While keeping the bladder as empty as possible during sleep, which lasts for about 10 hours, over distension of bladder is prevented, high pressure overflow incontinence ceases and by eliminating upper tract overfilling hydroureteronephrosis improves.

Overnight bladder drainage was well tolerated and was not associated with any significant adverse effects. The therapy for the valve bladder syndrome should be refocused to deal with the combined effects of polyuria, bladder insensitivity and residual urine volume.

In our study we found that the age at the time of initiating overnight bladder drainage and the time interval from initial presentation to the initiation of overnight bladder drainage played an important role in determining the ultimate outcome.

We are now initiating it in patients at a much younger age, not only to prevent the development of hydroureteronephrosis and the valve bladder syndrome but also to preserve renal function by eliminating bladder over distension.

Nocturnal bladder emptying is also ideally suited and currently used for the other causes of impaired bladder emptying especially neurogenic bladder. Preliminary results in this patient population have also been excellent.

While it will take long-term follow up to prove that this form of aggressive management preserves or at least delays the onset of renal failure, we believe that it is intuitively correct. Ultimately it is our hope that early intervention for residual bladder dysfunction will over time obviate the need for later augmentation and/or CIC programs in patients with posterior urethral valve altogether.

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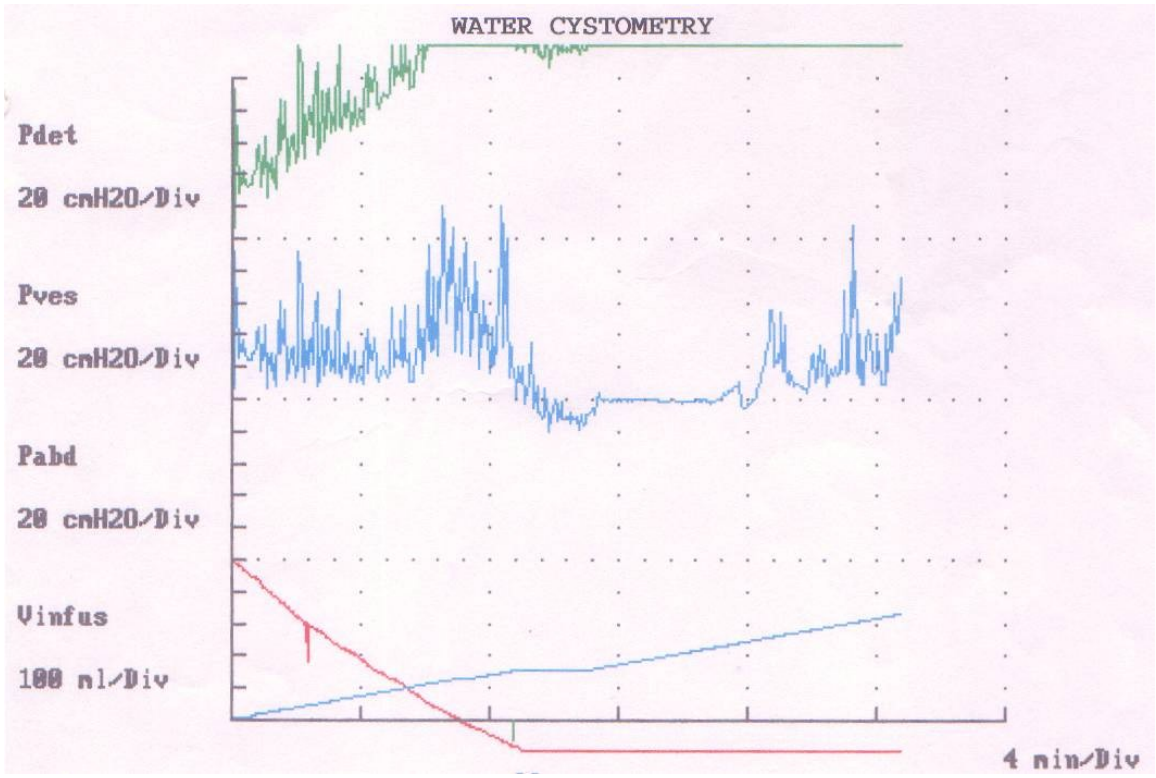
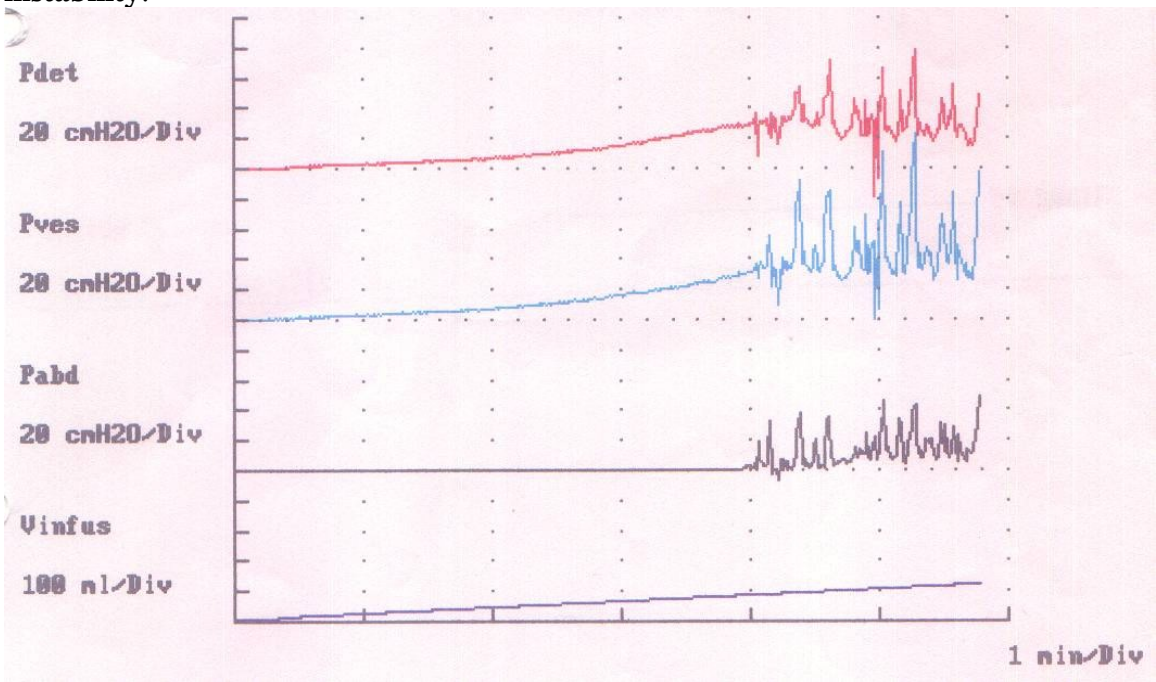
Cystometrograms of 10 cases Pre- and Post-OBD Stages

CASE-2:

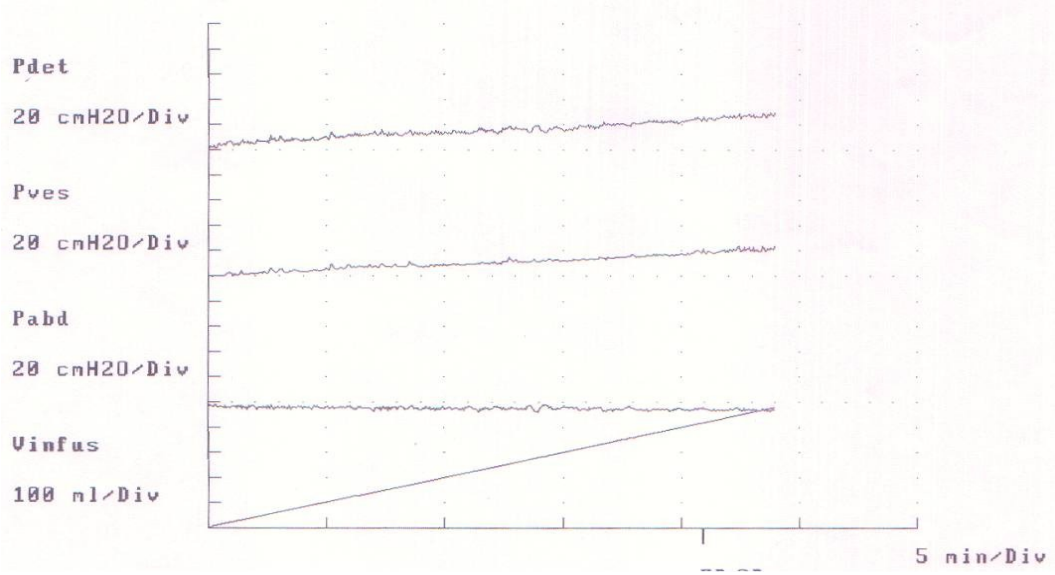


CMG at Pre-OBD stage showing small and Poorly compliant bladder with detrusor instability.

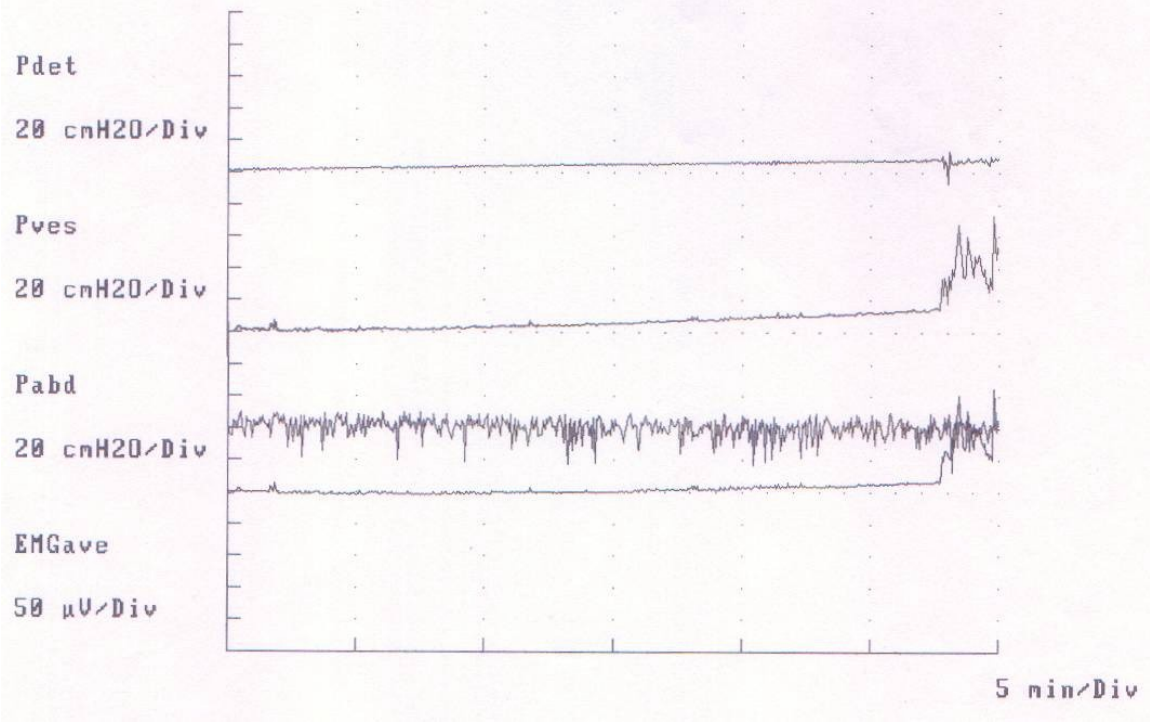


CMG at Post-OBD stage showing Low compliance bladder without detrusor instability.

Case-4

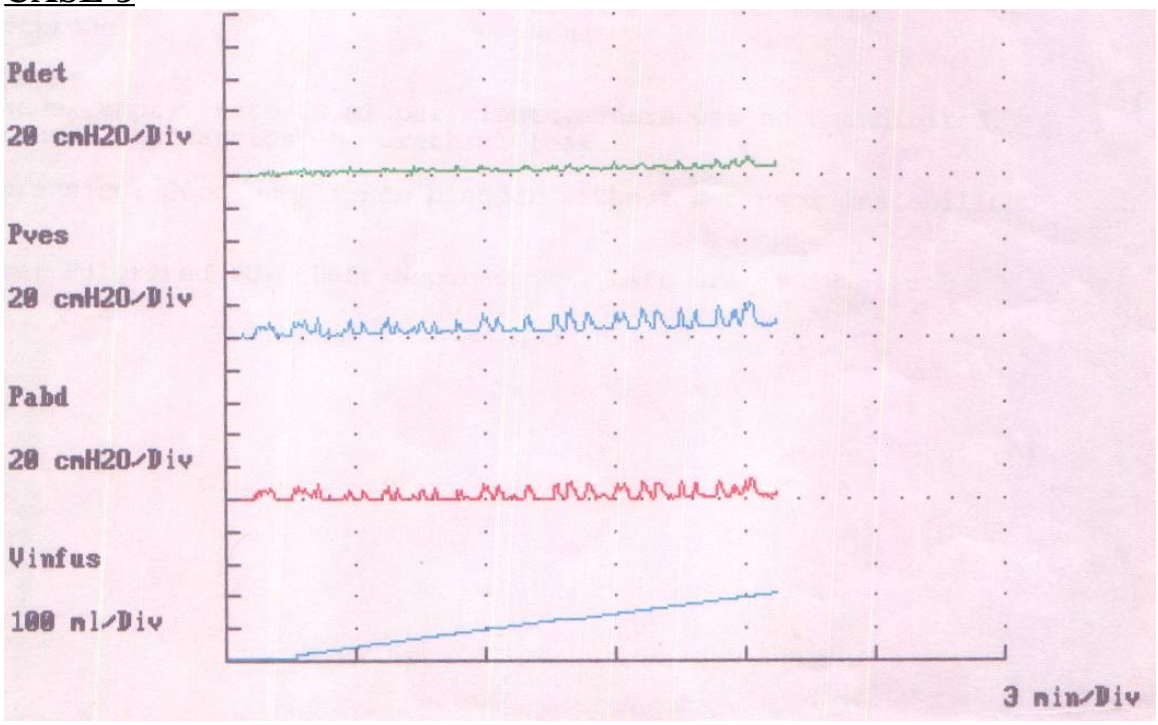


CMG at Pre-OBD stage showing Poor compliance bladder without detrusor instability.

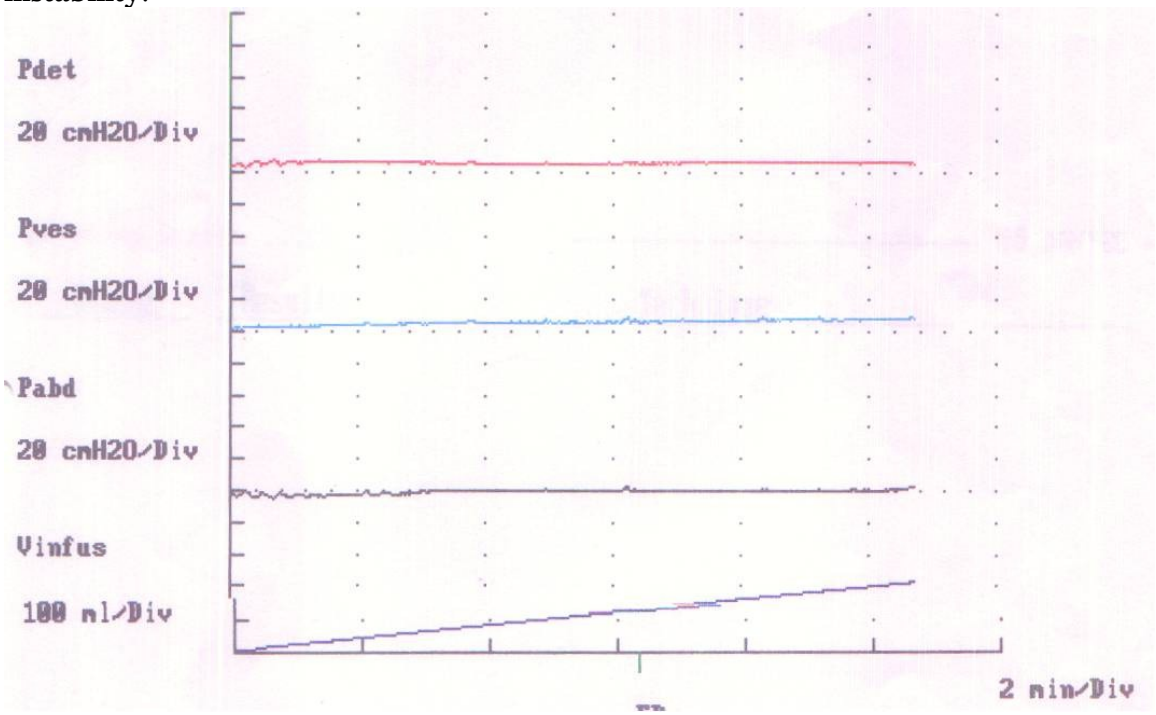


CMG at Post-OBD stage showing Good compliance bladder without detrusor instability.

CASE-5

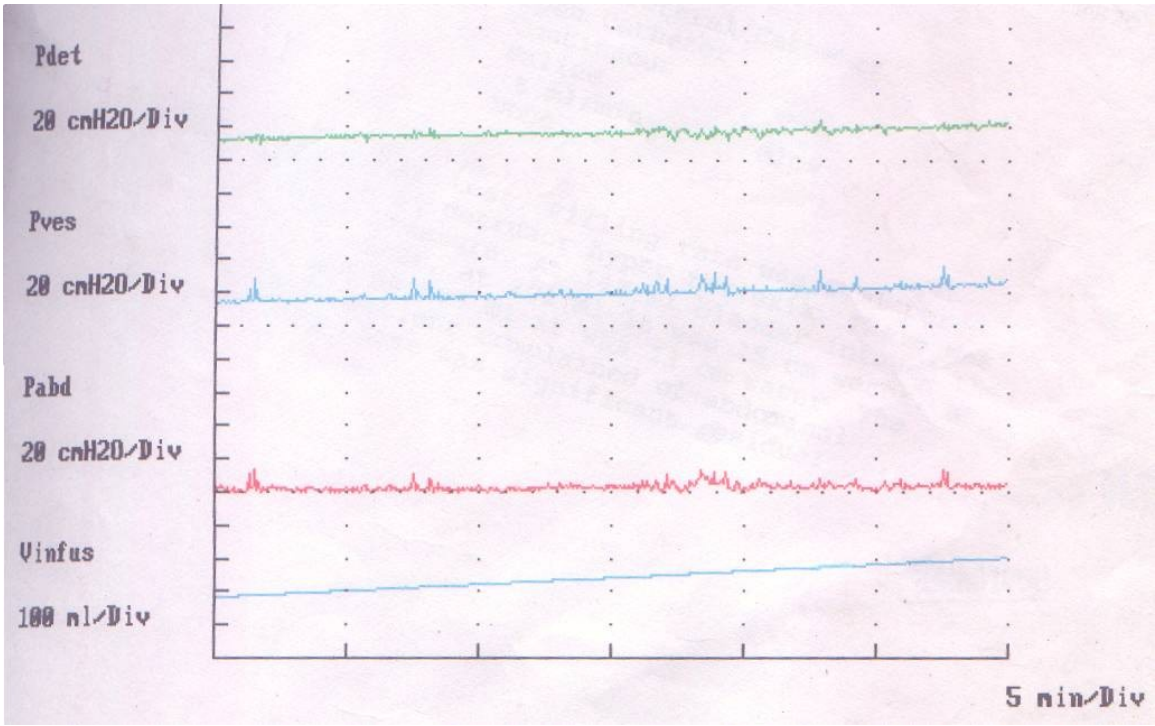


CMG at Pre-OBD stage showing Good compliance bladder without detrusor instability.

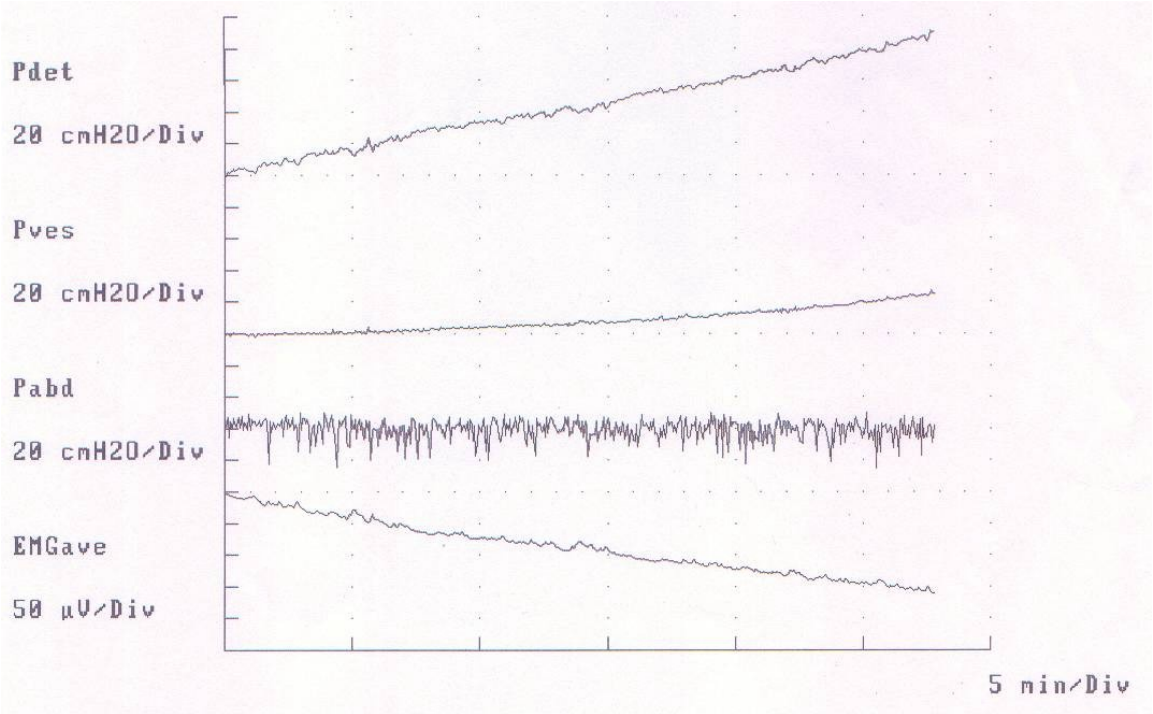


CMG at Post-OBD stage showing Good compliance bladder without detrusor instability.

CASE-7



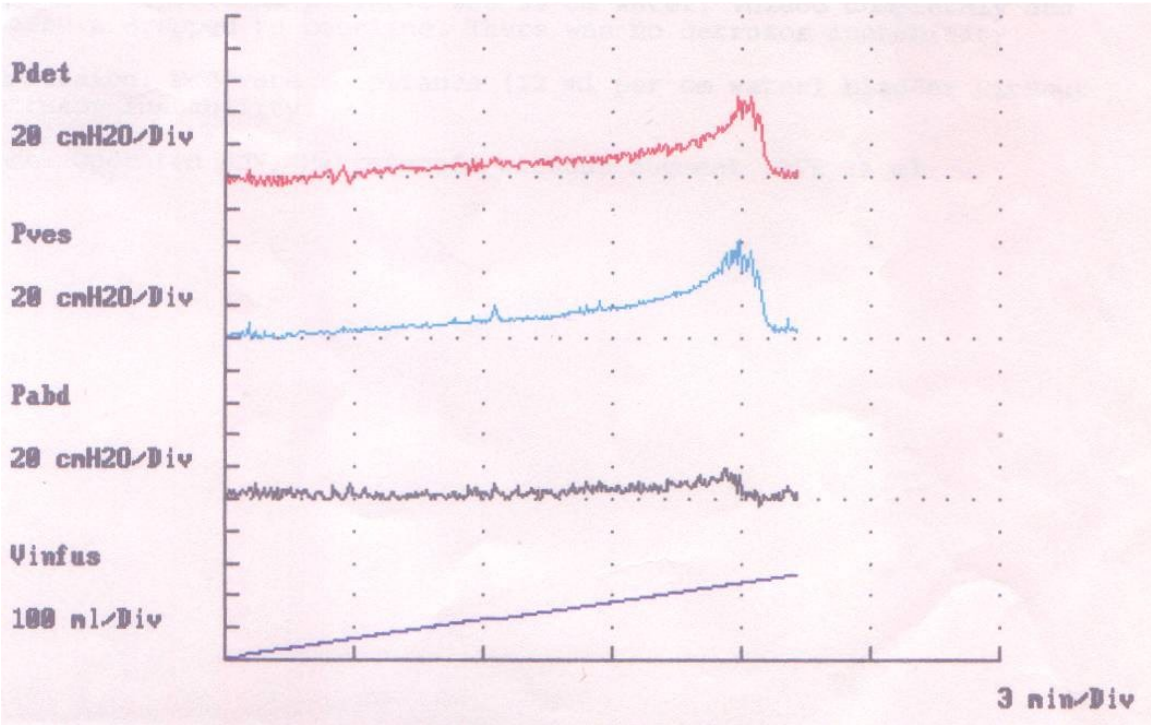
CMG at Pre-OBD stage showing Large capacity bladder with occasional detrusor instability, myogenic failure



CMG at Post-OBD stage showing Moderate compliance bladder without detrusor instability.

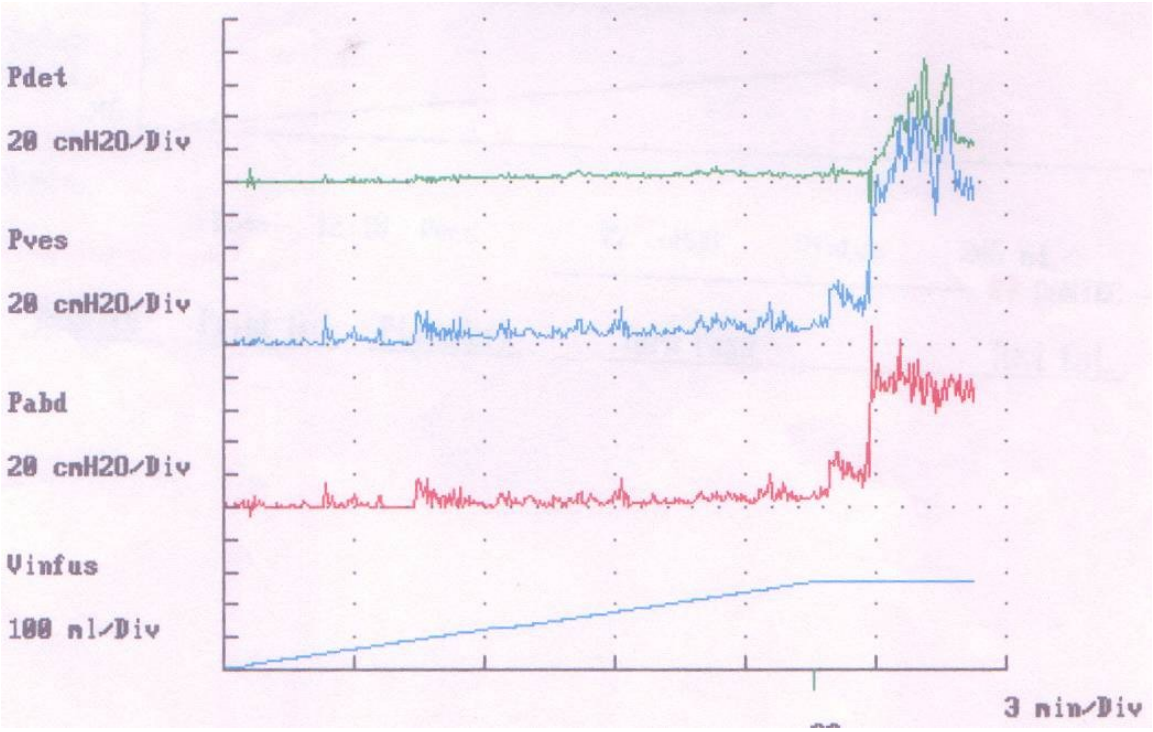
CASE-8

CMG at Pre-OBD stage showing Poor compliance bladder without detrusor instability.

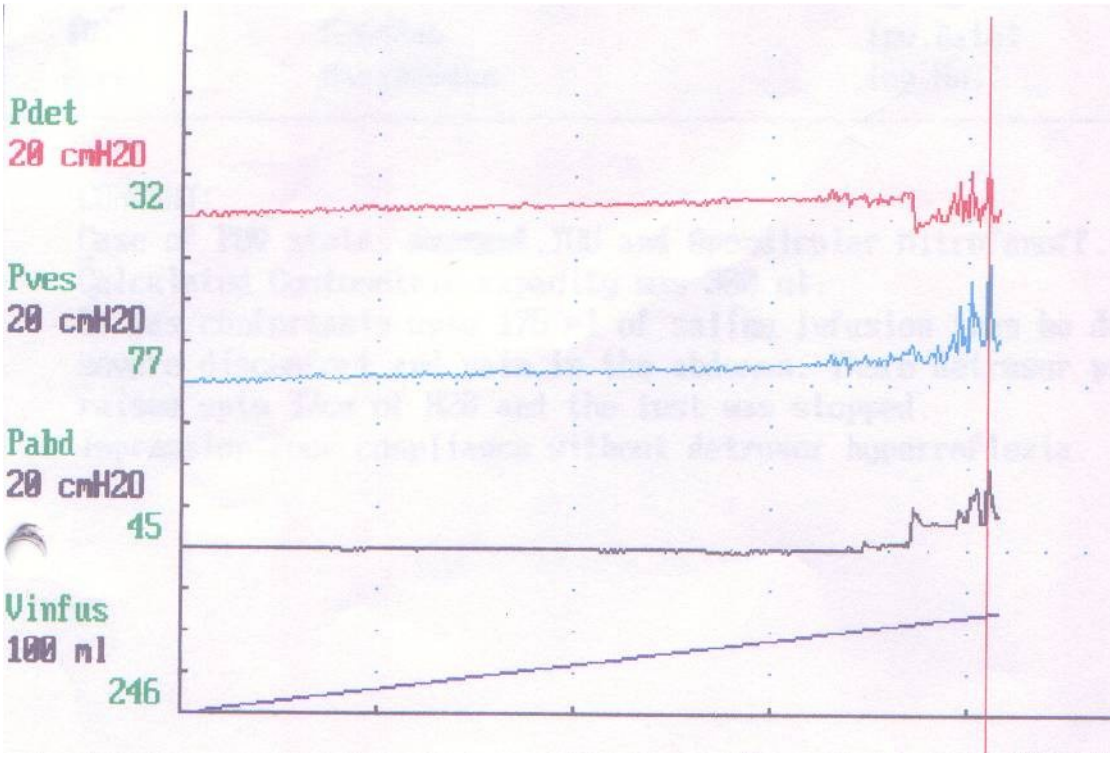


CMG at Post-OBD stage showing Moderate compliance small capacity bladder without detrusor instability.

CASE-9

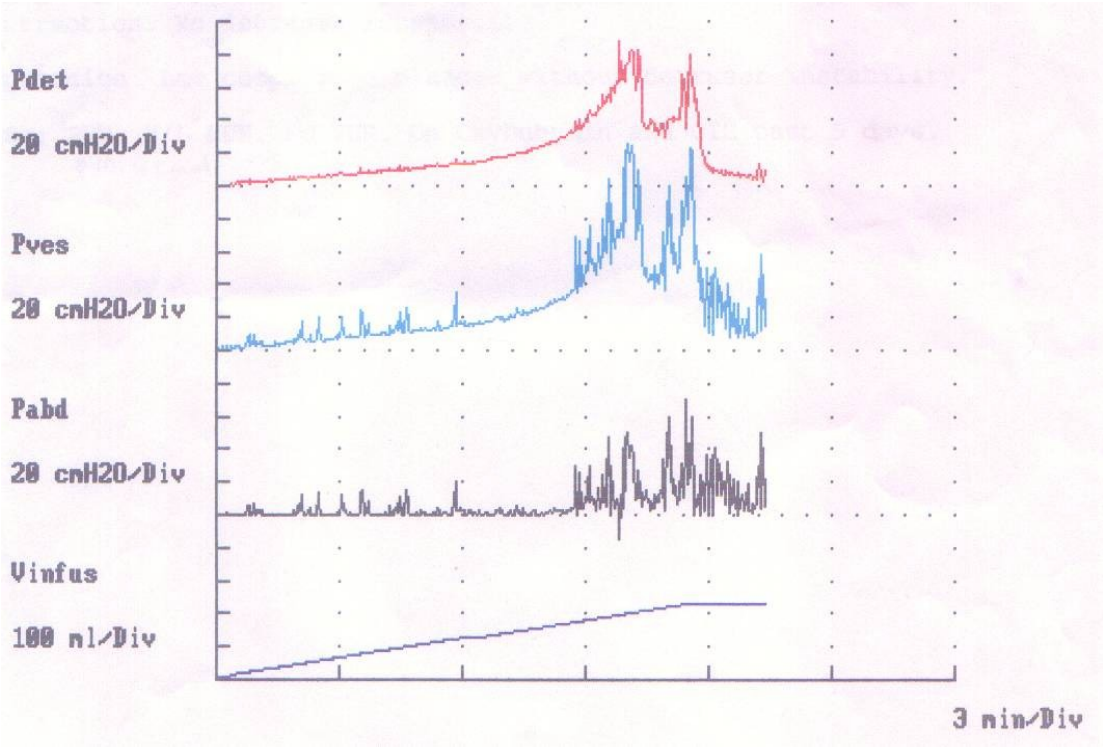


CMG at Pre-OBD stage showing Good compliance bladder without detrusor instability.

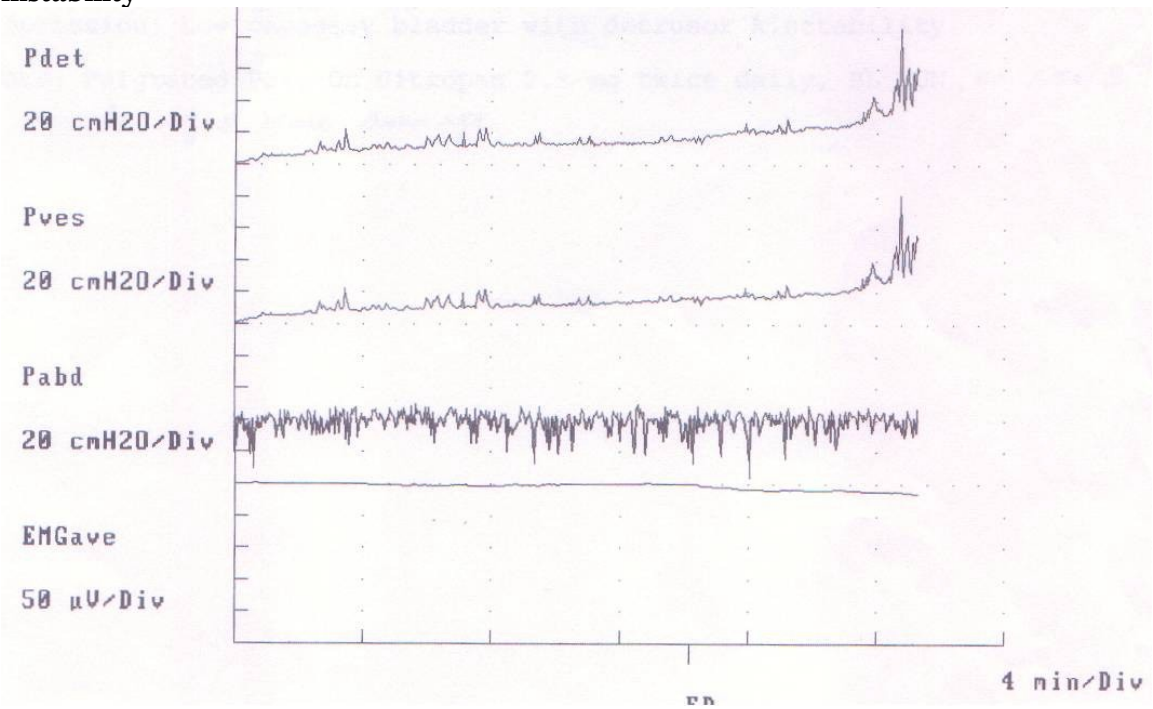


CMG at Post-OBD stage showing Poor compliance bladder without detrusor instability.

.**CASE-10**

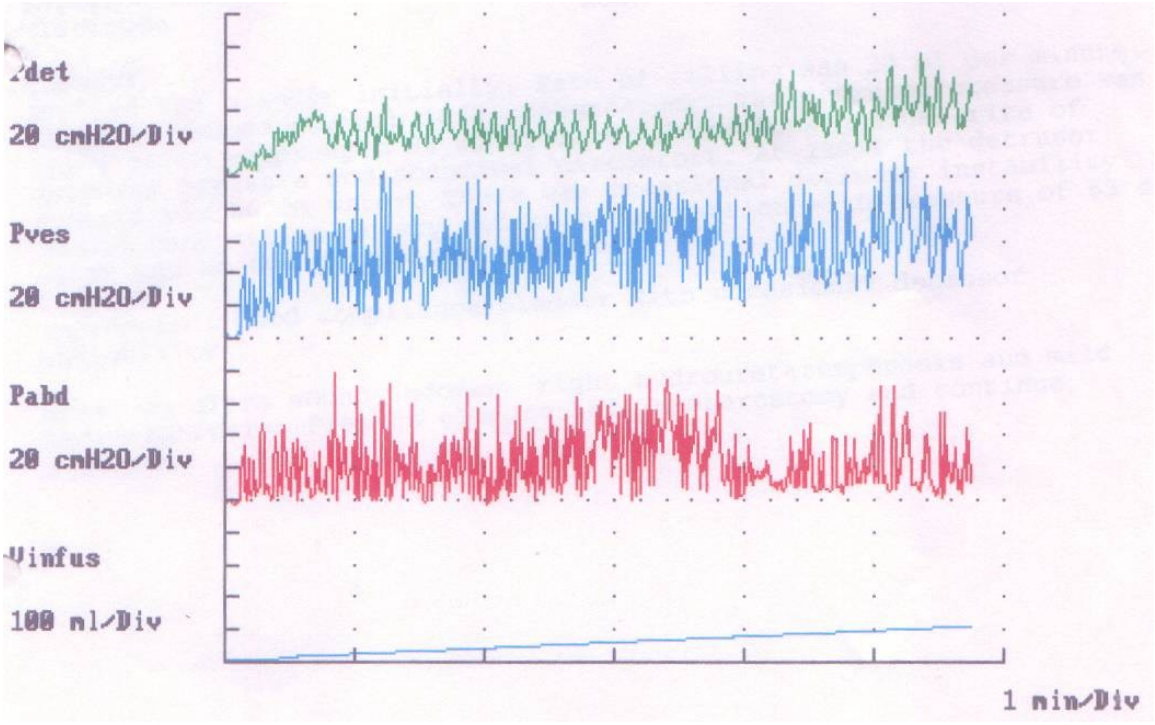


CMG at Pre-OBD stage showing Poor compliance bladder without detrusor instability

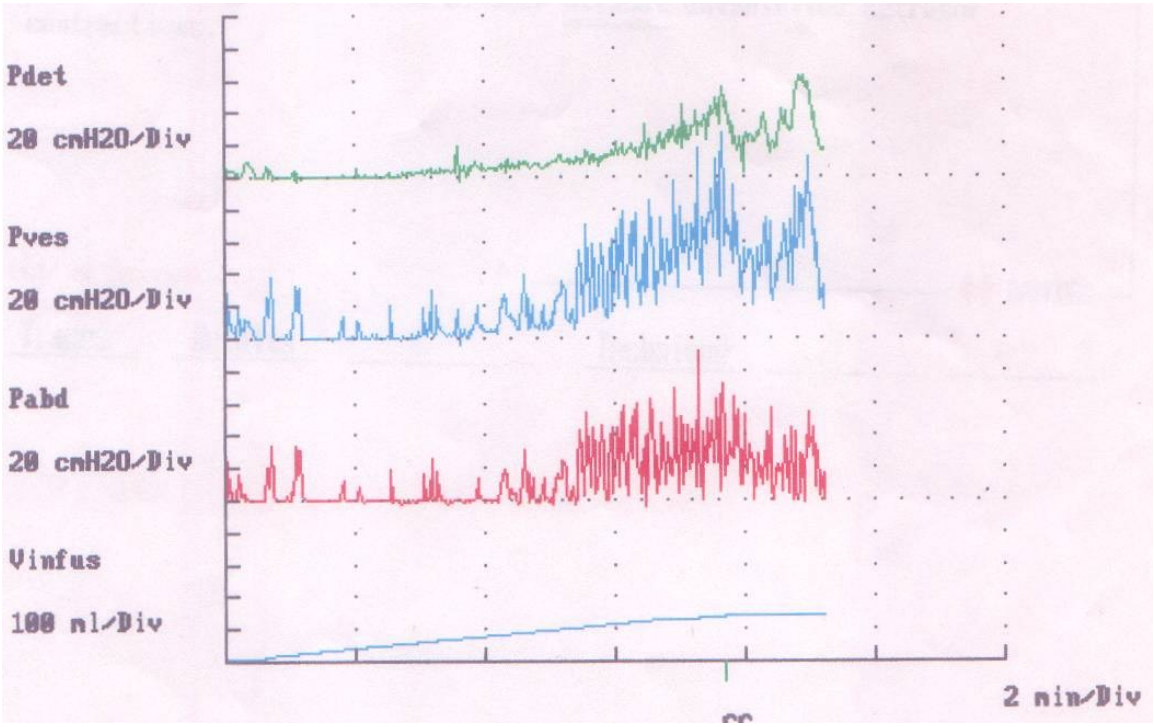


CMG at Post-OBD stage showing Poor compliance bladder with detrusor instability.

CASE-11

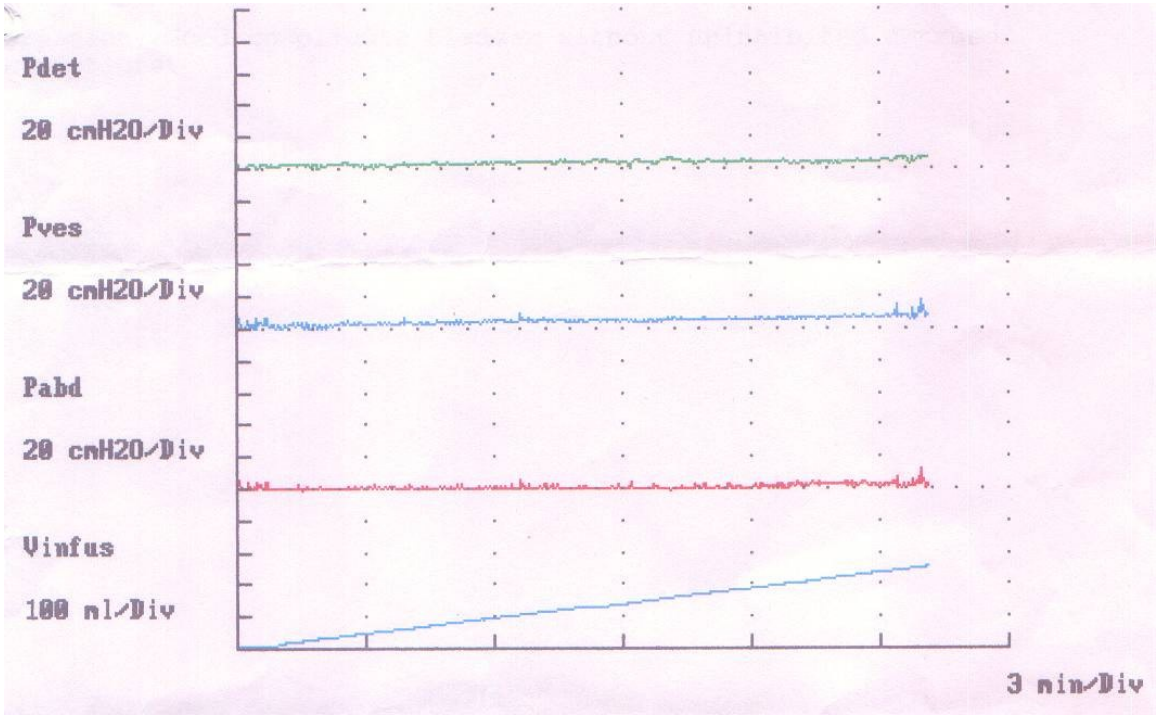


CMG at Pre-OBD stage showing Poor compliance bladder without detrusor instability.

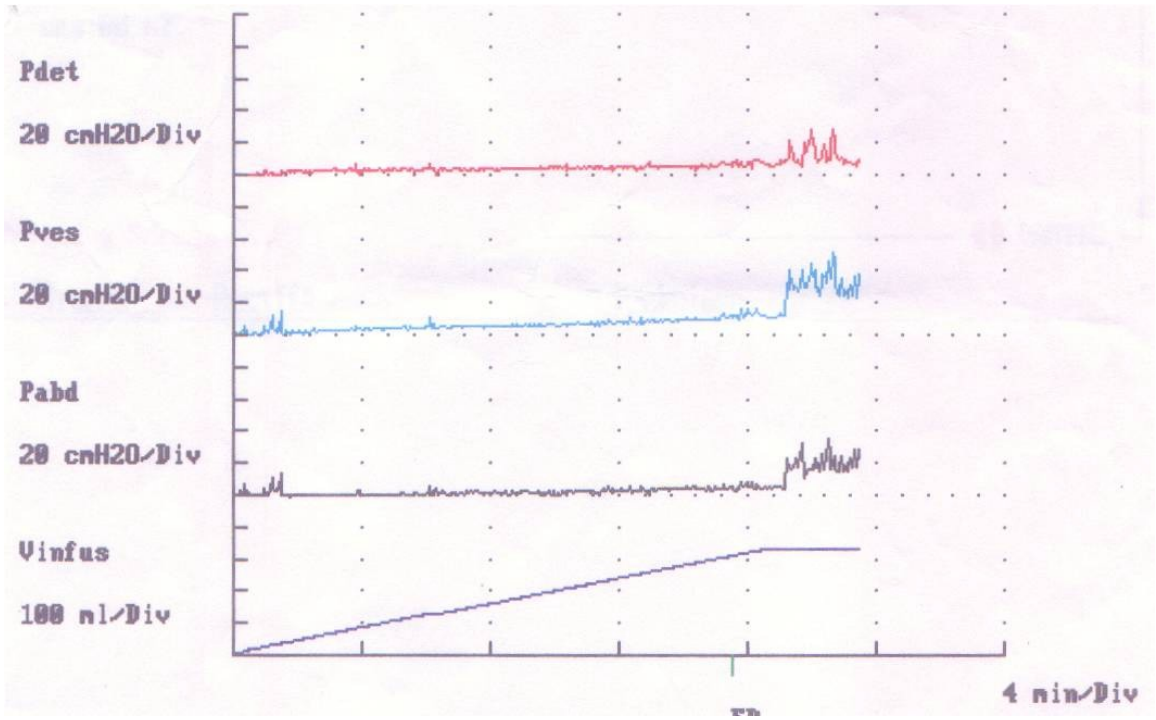


CMG at Post-OBD stage showing Good compliance bladder with occasional detrusor instability.

CASE-12

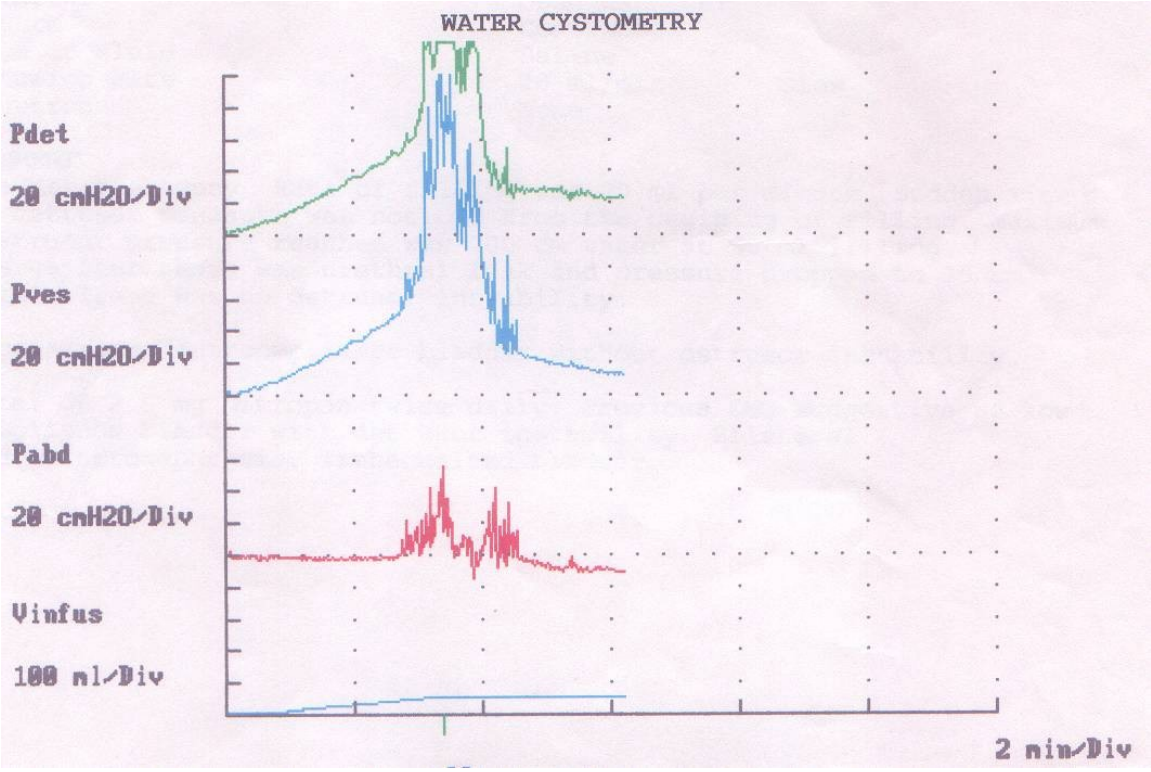


CMG at Pre-OBD stage showing Good compliance bladder without detrusor instability.

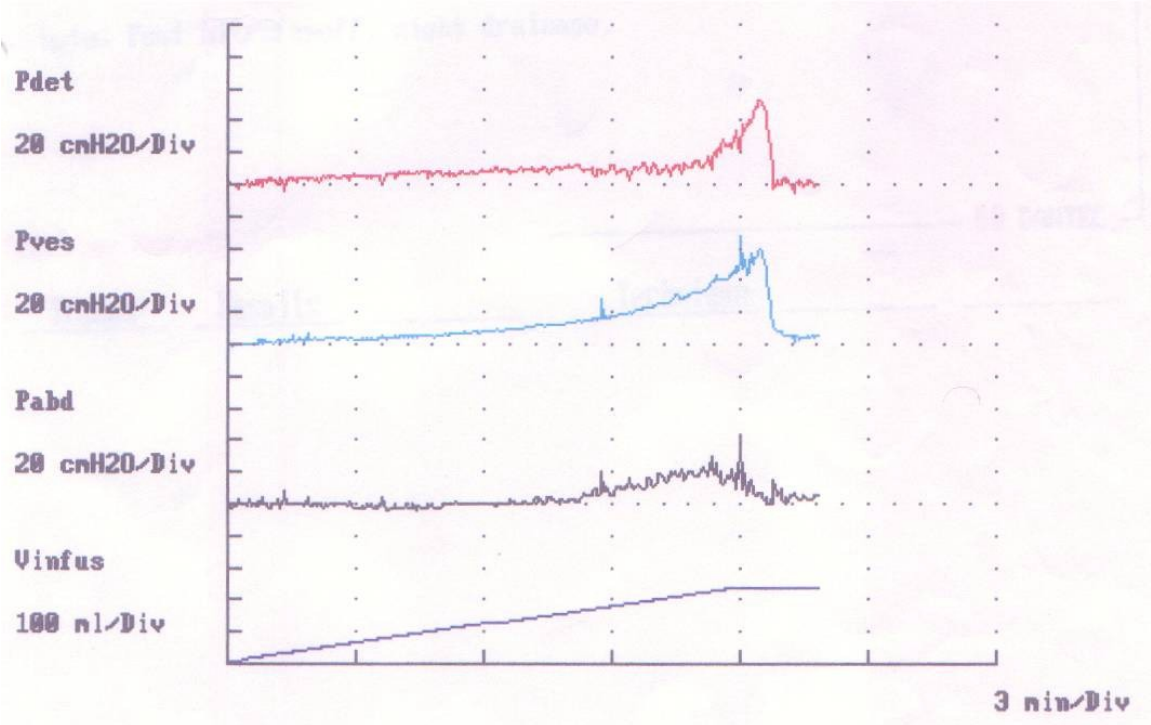


CMG at Post-OBD stage showing Good compliance bladder without detrusor instability.

CASE-15



CMG at Pre-OBD stage showing Poor compliance bladder without detrusor instability.



CMG at Post-OBD stage showing Good compliance bladder for 3/4th capacity without detrusor instability